NHS
The Leeds Teaching Hospitals
NHS Trust

Sickle cell disease in children & young people

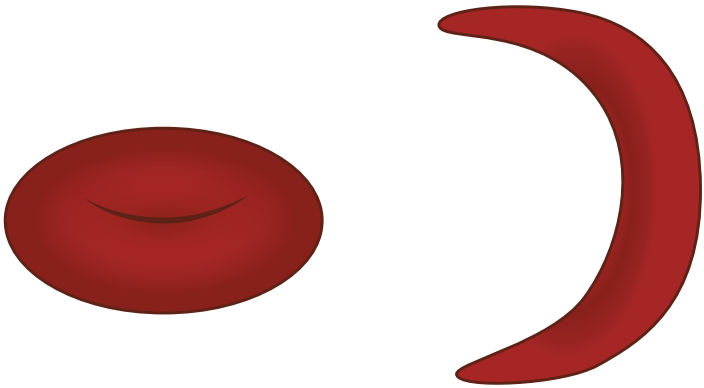
Information for patients & families

leeds children's hospital
caring about children

You have been referred to the Children's Haematology and Oncology Day Unit at the Children's Hospital at Leeds General Infirmary because you or your child has sickle cell disease. This booklet aims to give you some useful information about the unit and also some basic information about sickle cell disease.

What is sickle cell disease?

Sickle cell disease is an inherited disorder of the haemoglobin part of the red blood cell. Red blood cells carry oxygen around the body. In sickle cell disease the red blood cells can change into the shape of a farmer's sickle or crescent moon becoming sticky and rigid. When this happens the red blood cells can become stuck in the blood vessels causing a blockage. If a blockage occurs the blood cells can't deliver oxygen to that particular part of the body which causes pain and damage to the area. This is called **sickle cell crisis**.



Normal Cell *Sickle Cell*

Sickle cell crisis can be triggered by:

- Dehydration
- Not having your recommended medications and vaccinations
- Being too cold or too hot
- Excessive physical exertion/stress
- Emotional stress and anxiety

Information about the sickle cell clinic

Why do I need to come?

Sickle cell disease affects people in different ways and with variable severity. Some children have lots of severe pain and are often admitted to hospital. Others may experience mild pain or even in early years none at all. In these cases it is still really important to attend regular appointments as it is still possible the child could suffer from other problems, associated with sickle cell, affecting the heart, liver, brain or lungs and could still be at risk of stroke. If we know about these problems early we can offer treatment early to decrease the risk of you developing serious complications.

Some patients with sickle cell disease are on special treatments like hydroxycarbamide or receive regular blood transfusions, it is imperative that these are monitored closely in clinic.

Clinic times

General Children's Haematology Clinic

Thursday Afternoon: 2:30-4pm

In most cases your child will need to have bloods taken, the results will then be available when you see the doctor. For this reason **please be aware, the appointment time is for blood taking not the time you are due to see the doctor.**

There is at least a 30 minute wait after having blood taken, before you are taken through to see the doctor. We aim to see you within an hour of bloods being taken.

Sickle Cell Annual Review Clinic

Tuesday afternoon: 1st appointment 1:30pm - last appointment 4:30pm

All children and young people will be offered an annual review appointment. During these sessions you will see the Consultant Doctor and also the Clinical Nurse Specialist.

These sessions aim to give you a chance to ask questions and to help with your understanding of sickle cell disease. As your child gets older we will start to look at developing their understanding and look towards them gaining independence and moving to adult services when they leave full time education.

Who will I see at Clinic?

Receptionist

Please report to reception on arrival. You can contact reception on **0113 392 7179** to make or rearrange an existing appointment

Clinical Nurse Specialist

A nurse is always available to speak to if you have any questions or problems you want to discuss. You can also contact the Nurses on **0778 706 6296** or by email **benjamin.sykes@nhs.net**

Doctors

You will be seen by one of the Haematology Consultants (senior doctors specialising in blood disorders). **Or** one of the Haematology Speciality Registrars (Experienced doctors specialising in disorders of the blood).

Social Worker

Each child/young adult diagnosed with Sickle cell disease can be referred for social work support to help access social services and signpost to support around issues such as education, housing, benefits and immigration advice.

Nurses / Clinical support workers

You may be seen by a nurse to check blood pressure, pulse and oxygen saturations.

Play specialists

Play specialists use play as a distraction to help your child come to terms with medical procedures and their diagnosis.

Phlebotomists

The phlebotomist will take your blood sample.

For psychology, interpreter or spiritual support

Please contact the Clinical Nurse Specialist.

How often do I need to come to clinic?

- Children between the ages of 0-2 years will be seen at least every three months.
- Children between the ages of 2-5 years will be seen at least every six months.
- Children 5 years + will be seen at least annually.

Please note that we will always arrange to see your child if they are poorly, in pain, or you are concerned for whatever reason.

Please ring **0113 392 7379** between 08:30 - 18:00 to speak to a nurse on the day unit **or** If it is a bank holiday or outside day unit hours ring **0113 392 7431** to speak to a nurse on ward L31 the inpatient ward where we look after children with sickle cell disease.

You DO NOT need to go through Accident and Emergency (A&E)

What should I do if I can't come to clinic?

It is very important that you let us know if you cannot come to clinic. If you let us know that you cannot attend we can book someone else into your appointment time.

If you are unable to attend please call **0113 392 7179** between 09:00 - 16:30.

Situations in which you should contact the hospital straight away

Always contact us if you or your child has:

- Pain that is not controlled with pain medication
- Chest pain even if it gets better after pain killers
- Breathing becomes fast or laboured
- Swollen fingers or toes
- A temperature of 38°c or above
- Is pale, lethargic or shows any signs of anaemia
- Swollen tummy and pain on left side of the stomach
- Has a painful erection of the penis that does not stop
- Shows any sign of stroke:
  - **F** *- Facial weakness, can they smile? Has their face or eyes drooped?*
  - **A** *- Arm weakness can they raise both arms?*
  - **S** *- Slurred Speech*
  - **T** *- Time to ring the hospital, this is an emergency*

Contact:

The day unit (Mon-Fri 8:30am-6pm)
Tel: 0113 392 7379

Ward L31 (at any other time)
Tel: 0113 392 7431

You should NOT go through Accident & Emergency (A&E)

Guidance to help you care for your child if he/she has pain

When sickle cells get stuck in the small blood vessels they cause a blockage which prevents normal blood flow. These blockages cause pain wherever they occur often in the arms, legs, back and stomach, but can happen anywhere in the body. Sometimes this pain is quite severe. Blockages caused by sickle cells can also cause swelling of the hands and feet, stiff painful joints and extreme tiredness. Episodes of severe pain are known as "crises". Sickle pain can happen at home and at school - in fact anywhere - so it is important for children and their families, as well as carers and teachers, to know the best ways of coping.

Managing sickle cell pain at home

Mild sickle cell pain can be managed at home. In young babies and toddlers it may be difficult to know whether they are in pain or not. It is likely that you will notice that your child is not behaving as normal. They may be fretful and miserable, persistently crying, or only crying when moved. As your child gets older you will find that they get better at being able to tell you the site of the pain.

Following your first appointment with us, we will ask your GP to provide pain killers for your child. It is important to give pain medication as soon as your child complains of pain as it is then easier to control, rather than waiting until the pain is full blown and out of control.

Please make sure your child has plenty to drink and does not become dehydrated. Please make sure your child does not have a fever, if they do, contact the sickle cell centre. Please make sure they do not become cold.

We advise you use pain medication as follows:

Remember:
If your child has any of the symptoms described on the previous page contact the hospital for advice.

NB.
Only give medications as directed by pharmacist's instructions on the bottle or packet.

Use **Paracetamol** as soon as your child complains of pain

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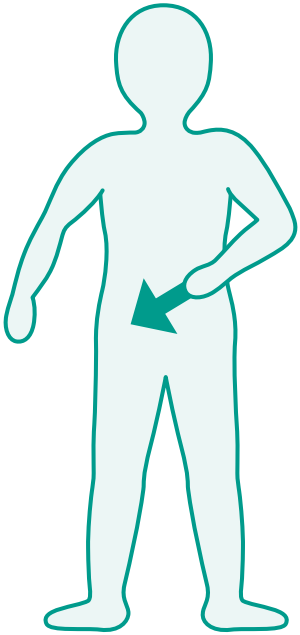
If still in pain after 30 minutes **add Ibuprofen** (Neurofen) but **continue to give Paracetamol** as directed

↓

If pain does not ease
Contact the day unit: 0113 392 7379 or
Ward L31 (if out of hours): 0113 392 7431

Feeling a large spleen

The spleen is a gland in the abdomen (tummy) that removes dead or dying red blood cells from the blood stream, it also helps protect the body from infections. It normally sits under the lower rib cage on the left side of the body. If it gets bigger it pushes into the abdomen down the left side and across to the lower right side, see diagram.



Patients with sickle cell disease often have small spleens but occasionally they can get very large. If they get bigger and push into the abdomen they can remove a large number of red blood cells from the blood stream which causes the body's red cell count or haemoglobin level to fall, a condition known as sickle cell splenic sequestration crisis. If the levels fall too low this can be a serious emergency and needs immediate attendance at the hospital and probably a blood transfusion.

It is therefore important that parents or carers can examine the abdomen of their child to feel if it is hard or swollen. Make sure your child is relaxed, perhaps after a bath or before bedtime. Place your hand at the bottom right of the abdomen and gently squeeze moving in small steps up to the top left, moving backwards from the point of the arrow in the diagram gradually up to its base. If the abdomen feels very firm or tender that may mean the spleen is enlarged. However if your child is not relaxed you may be feeling only tense muscles, it is not always straightforward. If you are worried there is a different feeling to normal and especially if your child is unwell or unusually pale, please ring the sickle cell centre telephone numbers immediately for advice.

Regular treatment for sickle cell

Penicillin V - People with sickle cell disease have difficulty fighting infections, particularly pneumococcal infection, and they risk becoming seriously ill. By taking penicillin V regularly, you can protect yourself from these infections.

The penicillin helps you to fight infection. It does not weaken their immune system.

You must give the penicillin twice every day to protect yourself, even if you think you are well.

Folic Acid - In patients with sickle cell disease, there are fewer red bloods cell than normal because sickled cells usually die after 10 to 20 days, in contrast to 120 days for normal red cells. The bone marrow, where the blood cells are made, has to work hard to keep making more cells. Folate is needed to make red blood cells and the body's stores of folate can run low.

Folic acid replenishes the depleted folate stores necessary to make more red blood cells.

Potential Treatment Options

Vitamin D - is needed to keep bones, teeth and muscles healthy and helps you grow. A lack of vitamin D can lead to bone deformities or bone pain. We get Vitamin D from sunlight so in the UK it is very normal to have low levels.

Hydroxycarbamide (also known as hydroxyurea) - is a medicine that is available as a liquid or a tablet and is used to reduce the frequency and intensity of sickle crisis.

How does it work?

In babies blood there is a special haemoglobin (Haemoglobin is the part of the blood that is affected by sickle cell) called haemoglobin F (HbF) which is not affected by a sickling process.

Hydroxycarbamide mainly works by reducing the amount of HbS made and increasing the amount of this baby blood HbF back towards the levels seen in young babies. This is very effective as HbF actually carries oxygen around the body and also prevents the sickle haemoglobin from sickling which is the fundamental process that causes almost all of the problems in sickle cell disease.

Hydroxycarbamide can reduce sickle symptoms and may protect against more serious sickle complications in the future.

If you want to know more about Hydroxycarbamide please speak to the doctor or specialist Nurse at your next appointment

Transfusions

If the sickle cell disease is causing significant harm we may need to use blood transfusions to dilute out the sickled red cells with healthy blood cells from a blood donor. This can either be

performed by a straight forward transfusion of blood into the veins – a 'top up transfusion' - or a process by which red cells are removed at the same time as being transfused - an 'exchange transfusion'. We may need to provide a top up or exchange blood transfusion as an emergency or on a regular basis. If regular transfusions are required we do this by performing red cell exchanges (usually every 6 weeks) we would aim to keep the sickle level below 30% (it is normally 80+%) and therefore greatly reduce the risk of sickle related problems. A significant advantage of the exchange procedure is that excess iron is not left in the body; regular top up transfusions will in time lead to excess iron in the body which can damage certain organs.

To be able to transfuse donor blood whilst taking out sickled red cells we need to put a cannula in both arms. Sometimes this isn't possible if the veins are not big enough, in these cases a vascath may need to be put in. A **vascath** is a catheter (flexible plastic tube) that is surgically inserted into a vein located either in the neck or groin.

If you want to know more about transfusion, please speak to the Doctor or Specialist Nurse at your next appointment.

Stem cell Transplant (sometimes called Bone marrow transplant)

Stem cell transplants are the only cure for sickle cell, but unfortunately they are not something that is suitable for everyone due to the significant risks involved. If you would like to know more speak to your consultant at your next visit.

What are stem cells?

Stem cells are produced in bone marrow (the spongy tissue found in the centre of some bones) and have the ability to develop into different types of blood cells.

For a stem cell transplant, the patient receives strong chemotherapy to kill their own bone marrow cells then stem cells from a healthy donor are given through a drip into a vein. These cells then start to produce healthy red blood cells to replace the cells containing sickle haemoglobin.

Will my child be able to have a stem cell transplant?

We will be happy to discuss stem cell transplantation with all families affected by sickle cell disease. Those patients with significant sickle cell related symptoms or complications will be most likely to benefit from this treatment. We would only be able to proceed if the patient is not affected by other significant health problems and if we can identify a suitable stem cell donor.

Travel Advice

Most people who suffer from sickle cell are able to travel abroad providing they take appropriate precautions.

Discuss travel plans with your GP or Consultant as early as possible (at least four weeks before you travel) so they can confirm you/your child are well enough to undertake the trip and advise on any necessary precautions.

When making a booking, make sure the travel agent, airline and insurance company are all aware of you or your child's diagnosis and medical condition.

If you or your child are unwell the day before or on the day of travel, consult your Consultant who will advise whether it is safe to travel.

What to take

A copy of a recent outpatient clinic letter - This should include your usual haemoglobin level and medications, any drugs you are allergic to and who to contact for advice in an emergency. If you are taking controlled drugs, for example morphine, make sure this is noted in the letter.

A letter to confirm that you are medically fit to travel - For insurance purposes.

Medication (including any painkillers you might need) - Make sure you have enough to last the whole trip. It is best to arrange this at least one week prior to travel. Take a copy of any prescription with you. For air travel all medicines should be packed in your hand luggage.

Mosquito net and insect repellent - If travelling to an area where there is malaria.

Rehydration salt sachets - These can be purchased from your local pharmacy.

Vaccinations

Make sure you are up to date with routine immunisations and Hepatitis B vaccine. Check any necessary travel vaccines and arrange these through your GP practice or a travel clinic at least six weeks in advance. If you are visiting an area that requires Yellow Fever vaccination or any other live vaccines and are currently taking the drug Hydroxycarbamide please discuss this with your Consultant as it may not be recommended.

Malaria

It is vitally important to take effective precautions for travel to or stop over in a country where malaria occurs. Your GP can advise on the choice of antimalarial medication. This should be obtained from your GP or a travel clinic. Other precautions must also be taken. These include using insect repellent which contains DEET, wearing clothing that covers your arms and legs especially at dusk and using a mosquito net.

Diarrhoea

Diarrhoea is common among travellers abroad. If you or your child have sickle cell disease this can cause dehydration which can trigger a crisis. To prevent this it is advisable to carry a supply of oral rehydration salt sachets which can be purchased from your local pharmacy. If you or your child develop fever or notice blood in the stools you should seek immediate medical advice.

During the flight

Keep you or your child mobile, warm and well hydrated throughout the flight. You or your child should drink plenty of fluids but avoid alcohol. Carry warm clothing or use a blanket to prevent chilling - the cabin temperature is often cold. If you or your child are old enough wear flight socks to reduce the risk of thrombosis - these can be bought in most large pharmacy stores.

Fertility and pregnancy

Both men and women with sickle cell are usually able to reproduce.

Sickle cell anaemia can become more severe during pregnancy and pain episodes may occur more often. A pregnant woman

with sickle cell is at higher risk for preterm labour and having a low birth weight baby.

Proper prenatal care and careful monitoring can improve the likelihood of a healthy pregnancy so it is extremely important to speak to your Consultant or specialist Nurse as soon as you know you are pregnant.

Future pregnancies

If you're pregnant you should talk to your Doctor and midwife as soon as possible. You will then have the option of being fast-tracked for a diagnostic test to find out if your baby's affected by sickle cell disease.

Couples who have a child with sickle cell disease may wish to discuss options for a future pregnancy that enables couples to avoid passing that condition on to their children. Options include testing at 11/ 12 weeks of the pregnancy, collection of umbilical cord stem cells at delivery to assess if the cells could be a potential stem cell transplant match or Pre-implantation Genetic Diagnosis (PGD).

PGD helps potential parents prevent the birth of a child with a serious genetic condition. It involves the use of assisted reproductive technology (ART). Eggs are obtained from the mother and fertilised with sperm through in vitro fertilisation (IVF). Once fertilised, the embryos develop for 5-6 days and then a number of cells are removed from each embryo. The genetic material (DNA or chromosomes) within these cells of the embryo is then tested for the genetic or chromosomal abnormality. An unaffected embryo is then transferred to the mother's womb to implant. If successful, the procedure will result in pregnancy and the child should not be affected by the condition for which it was tested.

Admission to hospital

Sometimes it may be necessary to treat your child in hospital. Our inpatient unit consists of three wards that provide treatment and care for children and young adults, who are being investigated or treated for cancer as well as sickle cell disease and other blood disorders.

Ward L31 - For children between 0-13 years old.

Ward L32 - Bone Marrow Transplant Unit

Ward L33 - Teenage unit

Visitors are encouraged at any time up until 8pm

Who can stay?

Due to limited space only one person can stay with your child at night time. We do encourage parents or family members to stay if possible. We do understand that sometimes this is not possible. Please let staff know if no one will be staying with your child. It is not possible for anyone under the age of 16 years to stay with your child.

What do I need to bring to hospital?

If you think there is a possibility your child might be admitted to Hospital it is a good idea to bring:

- All your child's medication
- Change of clothes
- Towel
- Toothbrush/Toothpaste /Soap / shower gel
- Nappies, Nappy bags, wipes / formula etc.

Useful contacts

Children's Haematology and Oncology Day Unit

Level C, Clarendon wing, Leeds Children's Hospital at Leeds General Infirmary

Reception: 0113 392 7179 (To re-arrange, cancel or check appointments)

Nurses: 0113 392 7379 (For medical advice)

Unit Manager: Michelle Kite, 0113 392 8806

Ward L31

Level A, Clarendon Wing, LGI
Tel: 0113 392 7431

Ward L32

Level A, Clarendon Wing, LGI
Tel: 0113 392 7432

Ward L33

Level A, Clarendon Wing, LGI
Tel: 0113 392 7433

Clinical Nurse Specialist for Sickle cell and Thalassaemia

Ben Sykes
Tel: 0113 392 7179 or 0778 726 6096
Email: benjamin.sykes@nhs.net

Suzie Preston
Tel: 0777 522 8860
Email: suzanne.preston1@nhs.net

Social Worker

Tel: 0113 392 2504

National Haemoglobinopathy Registry

The National Haemoglobinopathy Registry (NHR) is a confidential database of patients with red cell disorders (mainly sickle cell disease and thalassaemia major) living in the UK. The aim of the registry is to improve treatment and services for people with red cell disorders. Information obtained from the NHR helps to secure funds to develop improvements in treatment and patient care in your treatment centre. The information is also essential for healthcare planning, identifying patient numbers and research into improved treatment. Patients cannot be identified from reports published by the NHR as they are presented anonymously.

http://nhr.mdsas.com/wp-content/uploads/2017/03/NHR-Patient-Information-Leaflet-reviewed-060317.pdf

NHS Choices (benefits and allowances)

www.nhs.uk/Planners/Yourhealth/Pages/Benefits.aspx

The UK National Screening Committee

www.screening.nhs.uk

The Sickle Cell Society

Tel: 0208 961 779
Helpline: 0800 001 5660
www.sicklecellsociety.org

Feedback

We appreciate your opinion, please let us know if there is something we can do to improve our service or fill in a comment slip and place in one of the suggestion boxes on the day unit.

Unit Manager: Michelle Kite, 0113 392 8806

Patient advice and Liaison Service (PALS)

The aim of PALS is to resolve any problem or concern quickly.

Tel: (0113) 206 6261 - **Available during normal working hours only.**

Tel: (0113) 206 7168 - **For queries outside of normal working hours, please leave a voicemail**

Fax: (0113) 206 6146

Email: patientexperience.leedsth@nhs.net

We do value concerns and complaints as a means of improving the quality of our service and staff within the Patient Experience Department will be happy to guide you through the process, or answer any questions you may have.

As a point of information, if you are solely seeking compensation we are unable to deal with such requests under the NHS complaints procedure.

What did you think of your care?

Scan the QR code or visit **bit.ly/nhsleedsfft**

Your views matter

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Produced by: Medical Illustration Services • MID code: 20220304_006/BP

LN004302
Publication date 03/2022
Review date 03/2024