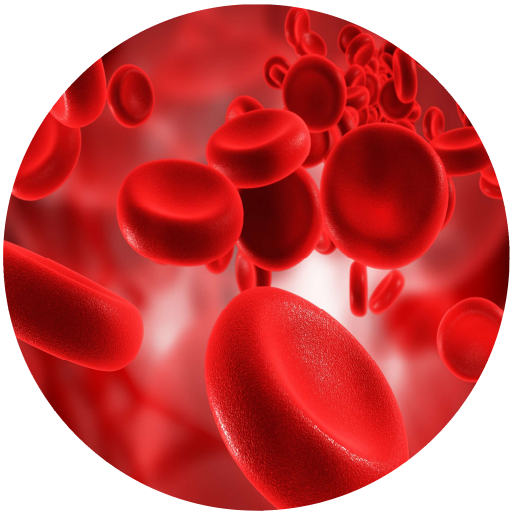

Information for patients with sickle cell disease who may need a blood transfusion

Patient information

This information leaflet answers some of the questions you may have about having a blood transfusion as part of your, or your child's, treatment for sickle cell disease.

All our leaflets can be accessed via the QR code at the end of this leaflet, including those designed for young children.

INF 1308/3

Effective: 08/02/2023 Review Date: 08/02/26

What is blood?

Blood is made up of red blood cells, white blood cells and platelets and these are carried around your body by a straw-coloured liquid called plasma. These cells play a vital role. Red blood cells carry oxygen, white blood cells fight infection and platelets prevent bleeding and help blood to clot. The adult human body contains about five litres of blood. Blood cells are made in your bone marrow. Red blood cells usually last 120 days in the blood of a healthy person.

Red blood cells contain a protein called 'haemoglobin' (sometimes called Hb). The haemoglobin gives the blood its red colour. Haemoglobin carries oxygen from the lungs to the organs and tissues of the body and then carries carbon dioxide back to the lungs where it is removed during breathing.

When there are not enough red blood cells in the body, either because the body is not making enough or because of bleeding, a person can become anaemic. Anaemia is the term used to describe a low haemoglobin level. Anaemia is picked up in a blood test called a Full Blood Count (or FBC), by measuring the haemoglobin level. A person is anaemic when their haemoglobin level is lower than the normal range for their age and sex. When you are anaemic, symptoms can include feeling tired, getting out of breath quickly, headaches, feeling dizzy and a pain in your chest. Anaemia affects people in different ways: for some, if it happens over a long time, they may not notice any symptoms until their haemoglobin level is quite low.

Normal red blood cells are round and flexible. In sickle cell disease, the red cells are sickle shaped which can make them rigid and sticky so they cannot bend and flex easily through the blood vessels. The red blood cells in sickle cell disease have sickle haemoglobin (HbS). These red blood cells only survive for about 10 –20 days in the body. The sickled red blood cells can block blood vessels which in turn can lead to severe, painful crises. This can also cause damage to organs such as the liver, kidney, lungs, heart, brain and spleen.

Why might I need a blood transfusion?

Many patients with sickle cell disease do not need a blood transfusion even if they have a low haemoglobin level.This is because their body becomes used to being anaemic and sickle haemoglobin releases oxygen more easily than normal haemoglobin. However, from time to time a blood transfusion may be needed, either because the anaemia has become worse or to lower the level of sickle haemoglobin in the blood.

Like all medical treatments, a blood transfusion should only be given if the benefits outweigh the risks. The healthcare team looking after you will discuss the specific reason for suggesting a blood transfusion and will explain the benefits and the risks to you.

There are two ways of receiving a blood transfusion for sickle cell disease; a standard transfusion (also known as a 'top-up' transfusion) or an exchange transfusion.

Why might I need a top-up transfusion?

Sometimes, anaemia in sickle cell disease can get much worse. You may need a 'top-up transfusion' to bring your haemoglobin up to a safer level and improve the oxygen supply to your tissues and organs.

You may need this:

- During a sickle cell crisis when there is a noticeable drop in your baseline haemoglobin level
- To reduce complications to you and your baby if you are pregnant
- Prior to surgery to reduce complications from the anaesthetic and the surgery.

Why might I need an exchange transfusion?

An exchange transfusion replaces your blood, containing sickle haemoglobin, with non-sickle blood from a blood donor. Exchange transfusion is used to stop or prevent damage to your organs by sickled red blood cells.

An exchange transfusion may be needed in an emergency such as:

- A crisis affecting the lungs (a chest crisis) or the brain (an acute stroke)
- A planned exchange transfusion that has been recommended before major surgery.

In addition, a series of regular planned exchange or top-up transfusions may be needed:

- As part of a long term transfusion programme to prevent stroke or other sickle complications
- In pregnancy.

Sometimes the blood can be removed and replaced with donor blood through a drip in the arm by a doctor or nurse – this is called a *manual* exchange transfusion. In some hospitals an *apheresis* machine is used where a patient is connected to a machine which is programmed to remove sickle blood and replace with donor blood, this is called an *automated* exchange transfusion. An automated exchange can sometimes only be performed during normal working hours and so the type of exchange transfusion performed can depend on both the hospital and the time of day.

How will my blood transfusion be given?

A bag of blood comes from one blood donor and is called a 'unit' of blood. A top-up transfusion is when blood is given through a small tube (a 'cannula' or 'drip'), usually placed in a vein in your arm. A unit of blood is generally transfused over 2-3 hours (maximum 4 hours from leaving storage). The number of units transfused depends on your size and the level of your haemoglobin.

For an exchange transfusion, blood will usually be removed from one vein and replaced through a tube in another vein at the same time. More units of blood are given with an exchange transfusion compared to a top-up transfusion and the blood is given more quickly. If your blood vessels are small, a special tube may be needed in the neck or the groin . These are known as central venous catheters (CVC) or 'lines'. This involves a small procedure, and usually these are put in place just before each transfusion and removed shortly afterwards. Occasionally a small device called a port can be inserted under the skin on the chest in a small operation. This allows easier access to a vein and avoids the need to have a new line for every transfusion. You should discuss the different options with your healthcare team.

Your healthcare team will be able to tell you where blood transfusions take place in your hospital and explain what facilities are available.

How will I feel during my blood transfusion?

Most people do not feel anything unusual during a blood transfusion. You will be observed before, during and after your blood transfusion. If you feel unwell during or after it you should inform your healthcare team immediately. Examples of feeling unwell are feeling dizzy, hot, or tired.

Do I need special blood?

All patients who receive a planned blood transfusion will have blood that is matched to their own blood group (ABO and D type), depending on what common molecules (antigens) are on the outside of their own red blood cells. Sometimes, because of small differences in the blood they receive, the body recognises the new blood as different to its own, and to protect itself, makes a protein known as an antibody. This means that if the patient needs a blood transfusion again, the hospital transfusion laboratory will need to perform additional tests to find blood that their antibody will not react with.

People with sickle cell disease need specially matched blood to reduce the risk of forming antibodies. The hospital transfusion laboratory needs to know that you have sickle cell disease to help provide this for you. This is particularly important if you are being treated somewhere other than your usual hospital. Please tell the healthcare team looking after you that you have sickle cell disease so that they can inform the transfusion laboratory.

Before every blood transfusion, your blood will be tested to confirm your blood group and check if there are any new red cell antibodies. Antibodies can develop in anyone, but are particularly common in people with sickle cell disease partly because of the number of transfusions they may have. You may be given a red cell antibody card if you do develop an antibody. This card will tell you which antibody you have developed. You should carry this red cell antibody card with you at all times, for example, you may want to keep a copy of this on a mobile device. Your healthcare team should ask to see your red cell antibody card but if they don't, tell them about it.

The transfusion laboratory will select blood for you that does not contain sickle haemoglobin and that is compatible with your blood type. The transfusion laboratory will attach a tag with your identification details to the unit of blood confirming that the unit is compatible for you.

Ask the doctors and nurses to check that the transfusion laboratory knows:

- That you have sickle cell disease
- If you have a red cell antibody card
- Where else you have had transfusions
- Whether you have had any transfusion reactions before
- If you think you may be pregnant.

What are the possible complications of blood transfusions?

Blood transfusions are common procedures that can save and improve lives. Most patients who receive a blood transfusion experience no complications or problems. Death due to transfusion is extremely rare. However, it is important that you inform a member of staff if you develop any symptoms during or after the transfusion.

Reactions

Some people may develop a temperature, chills or a rash during a transfusion. These reactions are usually mild and are easily treated with medicines such as paracetamol and antihistamines, and/or by slowing down or stopping the blood transfusion. Severe reactions to blood transfusion are extremely rare. An example of this is a 'haemolyic reaction' where the body destroys the blood that is being transfused. A patient having a severe reaction may be feeling very short of breath or very anxious. If a severe reaction does occur, staff are trained to recognise and treat them.

A build-up of fluid in the circulation during transfusion can cause breathlessness. You will be assessed before your transfusion to see if you are at risk, so that measures can be taken to prevent this. You should inform a member of staff immediately if you have any trouble breathing, so that treatment can be given at the earliest opportunity.

During exchange transfusion the anticoagulant (a blood thinner), which is added to each blood unit stop the blood forming clumps in the bag, can 'mop-up' the calcium in your blood and make you feel shaky or sick. Calcium supplements are given routinely in automated red cell exchange but extra can be given so tell a member of staff straight away if you have any symptoms.

Too much iron in your blood

This is common in people who receive repeated top-up blood transfusions though less so with automated red cell exchange. If you are on a transfusion programme, the iron level in your blood will be monitored. When necessary, excess iron can be removed by taking medication (injections or tablets).

Problems with cannulas or lines

In some people it can be difficult to find a vein for transfusion, particularly if you have had many transfusions before. Lines placed in the groin or neck may be uncomfortable and carry a small risk of causing bleeding, infection, or a blood clot. Your healthcare team will discuss this in detail if this happens to you.

Developing antibodies

Your blood is very carefully selected to match closely with the blood of the donor. However, it is still possible to develop antibodies against the donor blood and further screening is then needed to prevent a delayed haemolytic transfusion reaction. Having red cell antibodies can mean that matched blood may be harder to find. Some antibodies can become weaker or disappear so the transfusion laboratory may not know unless they are told about these. If you have developed red cell antibodies at any time in the past and you have been given a red cell antibody card to carry with you, show this to the healthcare team treating you.

Not having special blood

If you are being treated somewhere other than your usual hospital there is a small risk of not having specially matched blood if the transfusion laboratory don't know that you have sickle cell disease. It is important that you mention that you have sickle cell disease and that you need specially matched blood. If you have a red cell antibody card that says you need to have blood of a specific type, or if you know this from your medical history, please make a member of the healthcare team caring for you aware as soon as possible, and ask them to tell the hospital transfusion laboratory., When the transfusion laboratory know that you have sickle cell disease, they can also check on a a national computer system which helps them choose specially matched blood for you.

Delayed haemolytic transfusion reactions

A delayed haemolytic transfusion reaction is due to red cell antibodies reacting to the transfused blood. This can develop days or even weeks after the transfusion. These red cell antibodies can be newly-formed or may have been present before but too weak to be detected in the laboratory tests.

This may cause:

- Severe generalised sickle cell pain crisis
- Haemoglobin in the urine (red or 'cola' colour)
- Yellowing of your skin or the white part of your eyes
- Feeling noticeably tired and short of breath
- Fever
- Localised loin/back pain.

If you experience any of these symptoms, it is vital that you contact the hospital immediately.

You must inform staff that you have been recently transfused.

Are blood transfusions safe?

The risk that a blood transfusion will cause severe harm or even death is very low but this should be discussed with your healthcare team. In the United Kingdom we take many precautions to make sure any blood given to you is as safe as possible. One of the most important checks for a safe transfusion is to make sure you get the right blood. You can help reduce the small risk of being given the wrong blood by asking your healthcare team to check that it is the right bag for you.

You must be correctly identified at each stage of the transfusion to make sure that you get the right blood, including when blood samples are taken before the transfusion. **If you are an in-patient or a day case patient, wearing an identification band with your correct details is essential.** You will be asked to state your full name and date of birth and this will be checked against your identification band. If you know your NHS number, this is also useful to share. If you have your blood samples taken as an out-patient, you will not usually be given an identification band to wear, but it is still important that the staff ask you your full name and date of birth to confirm they are taking the samples from the right person. It is alright to remind the staff to ask you for this information and you can also ask to check that the sample taken from you has been labelled correctly before it is sent to the laboratory for testing.

Compared to other everyday risks, the likelihood of getting an infection from a blood transfusion is very low. All blood donors are unpaid volunteers and the risk of an infected unit entering the UK blood supply is extremely low. Donors and blood donations are tested for a number of infections which can be passed on through blood, but it is not practical or even possible to screen all donations for all infections, therefore, there will always be a small risk associated with having a blood transfusion.The risk of testing failing to detect a blood unit carrying a significant viral infection is less than 1 in 2 million (hepatitis B less than 1 in 2 million; Human immunodeficiency virus (HIV) less than 1 in 30 million; hepatitis C less than 1 in 60 million). It is strongly advised that all patients with sickle cell disease and thalassaemia are routinely vaccinated against hepatitis B.

Further information on the risks of transfusion can be found at: https://www.shotuk.org/patients/

Is a blood transfusion my only option?

You should be involved in all the decisions about your care and treatment. It is important that you understand the information and have the time to ask questions before you make your decision.

Your healthcare team will explain why you need a transfusion and will discuss the risks, benefits and if any alternative treatments are available. It is important you understand why a transfusion is required and that you have an opportunity to ask any questions.

Some people with sickle cell disease will never need a blood transfusion. In other situations, a blood transfusion can be life-saving and an important part of treatment**.** In an emergency, it may not have been possible to discuss all options at the time. If this happens, your healthcare team will talk to you about the blood transfusion you have had as soon as they can.

Giving your consent to transfusion

Before you give your consent to receive a transfusion, do you understand why you need the blood transfusion? And do you know the answers to these questions?

- What are the benefits of my transfusion?
- What are the risks of my transfusion?
- Are there alternative treatments I can try?
- What if I do nothing?

Please tell your healthcare team about any concerns you may have. It is important to share those worries or concerns; they will not think that these fears are trivial or of no importance. These may include:

- The impact on your other health problems
- The impact on future treatment options
- Religious and other non-health-related considerations
- Fear of needles, worries about feeling squeamish at the sight of blood or having had a bad experience in the past with a blood transfusion
- Your healthcare team having recommended special blood components based on several factors related to your treatment or your condition.

Once you understand what is involved and if you agree to have the transfusion, this consent will be recorded in your hospital notes. Sometimes, in an extreme emergency, you may not be well enough to have this conversation so your doctors may have to explain this to you when you feel better. If you have agreed to have a programme of exchange blood transfusions your consent will be reviewed periodically, particularly if anything changes.

As adults you have the right to refuse a blood transfusion, but you need to understand the consequences of doing so. Some medical treatments or operations cannot be safely carried out without a transfusion. In children, and patients who find it difficult to understand complex medical information, the medical team will work with the patient and their families or guardians, to make decisions that are in their best interests.

What if I have worries about receiving a blood transfusion?

If you are worried or have any questions, please talk to your healthcare team. Many hospitals have a dedicated Hospital Transfusion Team and, if appropriate, they may be able to come and discuss your concerns with you.

Acknowledgements:

We would like to acknowledge colleagues from Belfast Health & Social Care Trust, Croydon Health Services NHS Trust, Cardiff and Vale University Health Board, the National Blood Transfusion Committee Patient Involvement Working Group and the Transfusion Practice Panel who helped with writing this leaflet.

Additional Information

You may also find the following websites useful:

NHS :
https://www.nhs.uk/conditions/blood-transfusion/

NHS Blood and Transplant – blood website
https://www.blood.co.uk/the-donation-process/further-information/your-safety/
https://www.blood.co.uk/the-donation-process/

Sickle Cell Society website:

https://www.sicklecellsociety.org/

We would welcome your feedback and comments on this leaflet.

You can contact us in the following ways:

By post to:

Patient Blood Management

NHS Blood and Transplant

500 North Bristol Park

Northway

Filton

Bristol

BS34 7QH

By email to: PBM.team@nhsbt.nhs.uk

Or by phone: 01865 381010

This leaflet was prepared by NHS Blood and Transplant in collaboration with the National Blood Transfusion Committee. Further supplies can be obtained by accessing **https://hospital.nhsbtleaflets.co.uk**

Individual copies of this leaflet can be obtained by calling **01865 381010**.

The public can obtain the evidence sources for this leaflet by calling 01865 381010.

All of our leaflets can be accessed via this QR code

NHS Blood and Transplant saves and improves lives by providing a safe, reliable and efficient supply of blood and associated services to the NHS in England. We are the organ donor organisation for the UK and are responsible for matching and allocating donated organs. We rely on thousands of members of the public who voluntarily donate their blood, organs, tissues and stem cells.

For more information

Visit nhsbt.nhs.uk

Email enquiries@nhsbt.nhs.uk

Call 0300 123 23 23

INF 1308/3

Effective: 08/02/2023 Review Date: 08/02/26