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 treatment for your Sickle Cell Disease. You can also read the general NHS Blood and Transplant leaflet ‘Will I need a blood transfusion?’ for adults or information leaflets about transfusion designed for children and their parents.

**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. The adult human body contains about five litres of blood. Blood is made by your bone marrow, which constantly replaces the blood cells.

The haemoglobin (Hb) inside the red blood cells 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. Anaemia is the term used to describe a reduced number of red cells and a low Hb level.

People with Sickle Cell Disease have sickle haemoglobin (HbS) which can make red blood cells rigid and sickle-shaped so they cannot bend and flex easily through blood vessels like normal red blood cells. This can lead to small blood vessels getting blocked that 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 though they have anaemia as it is generally well tolerated. 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 it is essential. The doctors looking after you will discuss the specific reason for suggesting a blood transfusion for treatment of your Sickle Cell Disease and will explain the benefits and the risks to you.

There are two ways of receiving a blood transfusion for Sickle Cell Disease; a simple transfusion (also known as a ‘top-up’ transfusion) and exchange transfusion.

**Why might I need a top up transfusion?**

From time to time the anaemia in Sickle Cell Disease can get much worse. You may need a ‘top-up transfusion’ to bring the Hb up to a safer level and improve the oxygen supply to your tissues and organs.

You may need this:

- During a sickle crisis with a marked fall in haemoglobin level
- To reduce complications to you and your baby if you are pregnant
- Prior to major 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 (HbS), with normal blood from a blood donor. Exchange transfusion is used to stop, or prevent, a severe sickle cell crisis where there has been, or could be, damage to one of your organs by the 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 very severe painful crisis that is not improving with the usual medical treatment

A series of regular planned exchange or top-up transfusions may be needed:

- As part of a long term transfusion programme to prevent stroke
- In pregnancy or before surgery

Sometimes the blood can be removed and replaced through a drip in the arm by a doctor or nurse – this is called a manual exchange transfusion. Some hospitals have an apheresis machine that can be programmed to remove and replace the blood - this is called an automated exchange transfusion.

How will my blood transfusion be given?

Each bag of blood comes from one blood donor and is called a ‘unit’ of blood.

A top-up transfusion is where one or more units of blood are given through a small tube (a ‘cannula’ or ‘drip’), usually placed in a vein in your arm. Generally a unit of blood is transfused over 2-3 hours (maximum 4 hours). 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 arm and replaced through a tube in the other arm. More units of blood are given with an exchange transfusion compared to a top-up transfusion and the blood is given more quickly. When the exchange transfusion is automated, using an apheresis machine, a special tube may be needed in the neck (a central line) or the groin (a femoral line). Blood is stored in the fridge until needed and ideally should be warmed to blood temperature before exchange transfusion so that you do not get too cold during the procedure.

Your doctor or nurse 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 experience any unpleasant feelings whilst receiving a blood transfusion. To make sure you are alright, the nurses looking after you during the transfusion will periodically take your pulse, blood pressure and temperature and also check your breathing. If you feel unwell during or after a transfusion, you should inform someone immediately.
Nurses and doctors are trained to recognise any reactions to blood and to treat them. Occasionally you could experience minor reactions, which can be a bit worrying for you, but they are usually mild and easily treated. Severe reactions to blood are rare.

**Do I need special blood?**

People with Sickle Cell Disease need special blood and 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.

Every time you are being prepared for a blood transfusion, your blood will be tested to confirm the blood group and check if there are any red cell antibodies.

The transfusion laboratory will select blood that is ABO and RhD blood group compatible. Additionally, for those with Sickle Cell Disease, they will match for other Rh types (Cc Ee) and K. This matching reduces the risk of forming antibodies to these particular antigens present on the surface of red cells (though other red cell antibodies can still form).

The blood is then crossmatched to confirm that it is suitable to be transfused. 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 and
- If you have a red cell antibody card

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**What are the possible complications of blood transfusions?**

**Minor reactions**

Occasionally you may develop a temperature, chills or a rash during a transfusion. These are easily treated with paracetamol or an antihistamine.

During exchange transfusion the anticoagulant in donor blood, which is put there to stop the blood forming clumps in the bag, can ‘mop-up’ the calcium in your blood and make you feel a bit shaky. You may be given calcium supplements during the procedure if you need them.

**Iron overload**

This is common in people who receive repeated blood transfusions. If you are on a transfusion programme you will be monitored for iron overload. When necessary, excess iron can be removed by taking medication (injections or tablets).

**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 matching is then required 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 you should carry a card to alert anyone treating you.
**Delayed haemolytic transfusion reactions**

A delayed haemolytic transfusion reaction is due to red cell antibodies reacting to the transfused blood. These red cell antibodies can be newly-formed or may have been present before but too weak to be detected in the laboratory tests. The antibodies break down the blood within the two weeks of being transfused. This may cause:

- Severe generalised sickle cell pain crisis
- Blood in the urine (red or cola colour)
- Feeling tired and short of breath
- Fever
- Localised loin/back pain.

If you experience these symptoms contact the hospital immediately. You must inform staff that you have been recently transfused.

**Are blood transfusions safe?**

Yes, the risk that a blood transfusion will make you ill is very low. In the United Kingdom, we take many precautions to make sure any blood given to you is as safe as possible.

All blood donors are unpaid volunteers and there is a careful donor selection and testing procedure. It is not possible to screen for all diseases and there is always the risk despite our best efforts that you may acquire an infection. It is calculated that the risk of passing hepatitis B is very low at 1 in 2.2 million blood donations and it is strongly advised that all patients with Sickle Cell Disease on a transfusion programme are routinely vaccinated against hepatitis B.

The risk is many times smaller for HIV at 1 in 5.9 million and hepatitis C at 1 in 39 million (https://www.gov.uk/government/publications/safe-supplies-annual-review). The possibility of a blood transfusion transmitting variant Creutzfeldt-Jakob disease (vCJD – a rare, incurable brain disease) is extremely small and there have only ever been a handful of cases where patients are known to have become infected with vCJD from blood.

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 inpatient or 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. Please remind your nurse or doctor to ask you for this information if you think they haven’t checked properly!

**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 and make your decision.

If you are told that you might need a blood transfusion, you should ask your doctors to explain why it is necessary and whether there are any alternative treatments.

Many 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.
Giving your consent to transfusion

Once you understand what is involved and 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.

It is your choice to have a transfusion and you do have the right to refuse, but you should only do this after a full discussion with the doctors and nurses looking after you so that you are clear about how your decision may affect you. Other suitable treatments, if available, may be offered to you but you should be aware that some medical treatments or operations cannot be safely carried out without the option of a blood transfusion being given.

If you have any concerns you should discuss these with the doctors or nurses looking after you.

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We would like to acknowledge colleagues from St Thomas’, University College, Royal London and St Mary’s Hospitals and the Sickle Cell Society who helped with writing this leaflet.

Additional Information

If you are interested in finding out more about transfusion and have access to the Internet, you may find the following websites useful:

- **NHS Choices:**
  
  [www.nhs.uk/conditions/blood-transfusion/pages/introduction.aspx](http://www.nhs.uk/conditions/blood-transfusion/pages/introduction.aspx)

- **NHS Blood and Transplant – Blood Website**
  
  [http://www.blood.co.uk/about-blood/information-for-patients/](http://www.blood.co.uk/about-blood/information-for-patients/)

- **UK Transfusion Services**
  
  [www.transfusionguidelines.org.uk/index.aspx](http://www.transfusionguidelines.org.uk/index.aspx)

We welcome your feedback and comments on this leaflet.

You can contact us in the following ways:

**By post to:**

Customer Services, NHS Blood and Transplant, Part Academic Block – Level 2, John Radcliffe Hospital, Headley Way, Headington, Oxford OX3 9BQ

**By email to:** nhsbt.customerservice@nhsbt.nhs.uk

This leaflet was prepared by NHS Blood and Transplant in collaboration with the National Blood Transfusion Committee. Healthcare professionals can obtain further supplies by accessing [ww3.access-24.co.uk](http://ww3.access-24.co.uk) and entering their Regional Transfusion Committee code. If you do not have a code please call 01865 381010.

The public can get copies of this leaflet by calling 01865 381010.
NHS Blood and Transplant

NHS Blood and Transplant (NHSBT) saves and improves lives by providing a safe, reliable and efficient supply of blood and associated services to the NHS in England and North Wales. 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
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