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 treatment for 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 low Hb level.

People with Sickle Cell Disease have sickle haemoglobin (HbS) which can make red blood cells rigid, sticky and sickle-shaped so they cannot bend and flex easily through blood vessels like normal red blood cells. This can lead to 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 if 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 the benefits outweigh the risks. 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 standard transfusion (also known as a ‘top-up’ transfusion) and an 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 cell 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 problem caused or made worse by sickle cell disease 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 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 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 when blood is 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 vein and replaced through a tube in another vein. More units of blood are given with an exchange transfusion compared to a top-up transfusion and the blood is given more quickly. If the blood vessels are small, a special tube may be needed in the neck or the groin. These are known as central venous catheters (CVC). Blood is stored in the fridge until needed and ideally should be warmed to body temperature during 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 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 professional immediately.
Some people may develop a temperature, chills, a rash or breathing difficulties. These reactions are usually mild and are easily treated with medicines such as paracetamol and antihistamines, or by slowing down or stopping the blood transfusion. Severe reactions to blood transfusion are extremely rare. If they do occur, staff are trained to recognise and treat them.

**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 laboratory will then issue the blood for your transfusion. 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
What are the possible complications of blood transfusions?

**Minor reactions**
During exchange transfusion the anticoagulant in donor blood, which is added to stop the blood forming clumps in the bag, can ‘mop-up’ the calcium in your blood and make you feel a bit shaky. Calcium supplements are given routinely in automated red cell exchange and may be given in other forms of transfusion if you need them.

**Iron overload**
This is common in people who receive repeated blood transfusions though less so with automated red cell exchange. 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. 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?
The risk that a blood transfusion will cause severe harm or even death is very low but this should be discussed with your healthcare professional. 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 professional 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, 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 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 healthcare professional to ask you for this information.
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 continues to decrease\(^1\). Donors and blood donations are screened for a number of infections which can be transmitted 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.

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 and Thalassaemia are routinely vaccinated against hepatitis B.

The risk of getting variant Creutzfeldt-Jakob disease (vCJD) from a blood transfusion is extremely low. Each year, approximately 2.6 million blood components are transfused in the United Kingdom and there have been only a handful of cases where patients are known to have become infected with vCJD. More information on vCJD can be found on the NHS Choices website: [www.nhs.uk/conditions/Creutzfeldt-Jakob-disease/Pages/Introduction.aspx](http://www.nhs.uk/conditions/Creutzfeldt-Jakob-disease/Pages/Introduction.aspx)

Further information on the risks of transfusion can be found at: [www.shotuk.org/home/](http://www.shotuk.org/home/)

**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.

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.
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 need to fully understand the consequences of not doing so. 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 a blood transfusion.

What if I have worries about receiving a blood transfusion?

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

Acknowledgements:

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**Additional Information**

As a precautionary measure to reduce the risk of transmitting vCJD, people who have received a blood transfusion of blood or any blood component since 1980 are currently unable to donate blood or blood components.

You may also find the following websites useful:

**NHS Choices:**
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/

**Reference**

We would 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
Or by phone: 01865 381010
NHS Blood and Transplant (NHSBT) saves and improves lives by providing a safe and reliable supply of blood components, organs, stem cells, tissues and related services to the NHS and other UK health services.

We manage the UK wide voluntary donation system for blood, tissues, organs and stem cells, and turn these donations into products that can be used safely to save lives or radically improve the quality of people’s lives.

Any information that is passed on to NHSBT is held securely and the rights of these patients are protected under the Data Protection Act (1998).