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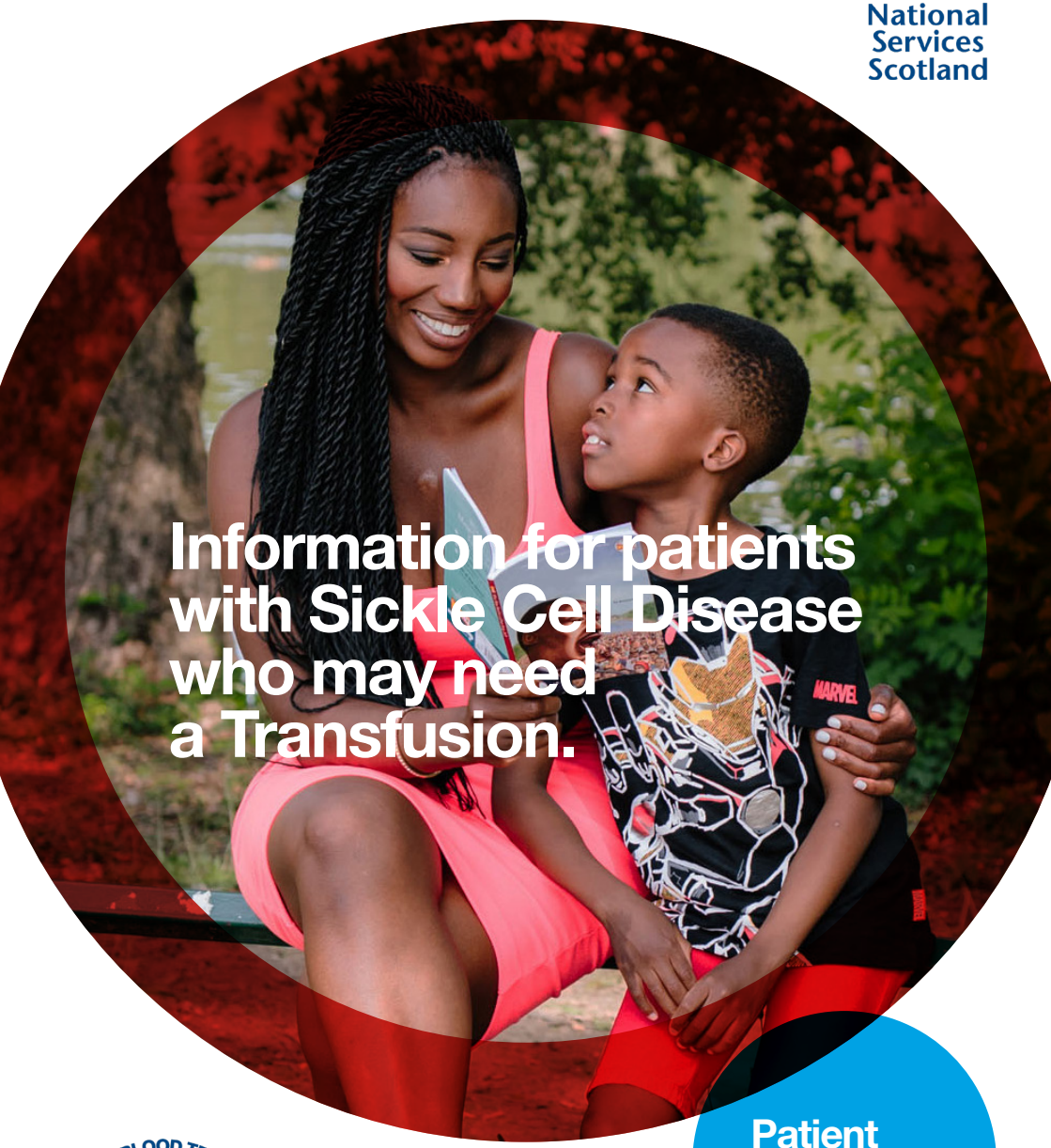

Information for patients with Sickle Cell Disease who may need a Transfusion.

Patient information leaflet.

SCOTTISH NATIONAL BLOOD TRANSFUSION SERVICE®

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 "Receiving a Blood Transfusion" patient information leaflet (PIL) which has been developed for adults and children. Your healthcare team will be able to provide you with a copy or alternatively you can download a copy from the following:

www.nss.nhs.scot/blood-tissues-and-cells/snbts-transfusion-team/national-policies-factsheets-and-patient-information/

What is blood and what is Anaemia?

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 and is common in people with Sickle Cell Disease. This low Hb level means that blood cells cannot carry enough oxygen around the body giving symptoms like fatigue and breathlessness.

What else happens in Sickle Cell Disease?

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 healthcare team 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, risks and alternatives 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)
- As part of a long term transfusion programme to prevent stroke or other sickle complications
- In pregnancy or before surgery

Exchange transfusions are performed through a machine, called a cell separator or apheresis machine, which can be programmed to remove and replace the red cells by separating the blood into its components using a centrifuge. In this process, only the red cells are removed and replaced by the donors' red cells and the rest of the blood is returned. This is called an ***automated*** exchange transfusion.

In rare circumstances the blood can be removed and replaced through a drip in the arm – this is called a ***manual*** 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 healthcare team will be able to tell you where blood transfusions take place in your hospital and explain what facilities are available.

If time allows you will be able to visit the apheresis unit where these automated red cell exchanges take place so you can become familiarised with staff and the apheresis machine. You will also be given the opportunity to ask questions about the exchange procedure.

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. Antibodies are a type of protective protein that can be produced by your immune system in response to a foreign material.

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 red cell antigens including K and other Rh types (Cc and Ee).

This matching reduces the risk of forming red cell antibodies to these particular antigens present on the surface of red cells, though other red cell antibodies can still form.

All efforts will be made to provide blood that has been screened for HbS and is negative.

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 your healthcare team 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?

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
- Dark colour urine (red or 'cola' colour)
- Worsening jaundice
- 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.

Citrate reactions

During automated red cell exchange transfusion, the anticoagulant used to stop the blood forming clumps, can 'mop-up' calcium in your blood and make you feel a bit shaky. To avoid this, calcium supplements are given routinely.

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 blood component 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.

Correct identification is crucial - Please feel comfortable in reminding the member of staff to ask you for this information if they do not do so.

Blood components are donated by healthy, unpaid volunteers and the risk of an infected unit getting into the UK blood supply is extremely low. Donors complete a health questionnaire every time they donate and blood donations are tested every time for a range of potential infections, including hepatitis B, C and E, and HIV. This makes the chance of transmitting any infection very low, but the risk can never be removed completely.

- The risk of testing failing to detect a blood unit carrying a significant viral infection is less than 1 in a million (Hepatitis B less than 1 in 1 million; HIV and Hepatitis C less than 1 in 10 million).

It is strongly advised that all patients with Sickle Cell Disease are routinely vaccinated against hepatitis B.

The chance of contracting variant Creutzfeldt-Jakob Disease (vCJD) from a transfusion is very small; nevertheless, we exclude donors who may be at a higher risk of vCJD. **For this reason, anyone who has received a blood transfusion or any other blood component since 1980 is currently unable to donate blood or blood components.**

For more information about how the risk of transfusion transmissible infectious agents entering the blood supply in UK is minimised, you can visit: www.transfusionguidelines.org/document-library/position-statements

- Bacteria could contaminate red cells and other components of blood. This could cause a dangerous reaction in any patients who receive contaminated units. We work hard to prevent this happening and the risks are now similar to the other infections listed above.

Further information on the hazards of transfusion can be found at: 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 healthcare team 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. More information on consenting to transfusion can be accessed at the following website:

Guidelines from the expert advisory committee on the Safety of Blood, Tissues and Organs (SaBTO) on patient consent for blood transfusion - GOV.UK (www.gov.uk)

What if I have worries about receiving a blood transfusion?

If you are worried or have any questions, please talk to your healthcare professional.

Duty of Candour

The Scottish National Blood Transfusion Service (SNBTS) complies with Duty of Candour legislation. This means we will act in an open and transparent manner where an unexpected or unintended event has occurred, which appears to have caused harm or death in direct relation to transfusion. Please ask your healthcare team for further information or access the following link:- www.gov.scot/policies/healthcare-standards/duty-of-candour/

Acknowledgements:

We would like to acknowledge NHSBT whose resources were used in producing this leaflet.

This leaflet has been reviewed and approved by the Scottish Paediatric and Adult Haemoglobinopathy Network (SPAH)

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

The Scottish Paediatric and Adult Haemoglobinopathy Network (SPAH) consists of healthcare workers in Scotland who have responsibility for the care of children and adults with haemoglobinopathies – mainly sickle cell disease and thalassaemia. SPAH is approved and supported by the National Service Division of NHS Scotland as a managed clinical network.
Further information about the network is available at www.spah.scot.nhs.uk

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

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