SICKLE CELL AWARENESS

sickle cell disease in the UK

Sickle Cell Disease (SCD) affects around

15,000

people in the UK

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 cells.

This can lead to small blood vessels getting blocked which:

- can lead to severe painful crises.
- can cause damage to organs such as the liver, kidney, lungs, heart, brain and spleen.

The Sickle Cell Society has produced the following information leaflets available at sicklecellsociety.org

- Let's talk about sickle cell
- Did You Know? Age 5-10
- Sickle Cell Disorder and Sickle Cell Trait

There are also guides for teachers, healthcare professionals and employers

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NHS Blood and Transplant

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when is blood transfusion needed?

People with SCD have anaemia which is generally well tolerated.
The majority of patients don't need transfusion.

However from time to time the anaemia can get much worse and a transfusion may be needed. This is known as a **"top up" or "simple" transfusion**

Sometimes an **"exchange transfusion"** may be needed to remove sickle blood and replace this with normal blood

This may be needed if there is a sickle crisis affecting an organ such as the lung ('chest crises') or the brain when transfusion is needed to prevent or treat stroke.

Testing children with SCD using ultrasound can help identify those at the risk of stroke - this risk can be reduced with a regular transfusion programme.

Blood transfusion may sometimes also be needed before surgery, during pregnancy or other situations.

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safe blood transfusion in sickle cell disease

There is an **increasing need** for blood transfusion in patients with Sickle Cell Disease

Patients with SCD have

'special' requirements

for blood.
In addition to ABO and D matching they should also receive blood that is matched for other Rh types (C, c, E, e) and K.
This reduces the risk of forming antibodies to these antigens present on the surface of red cells.

All patients needing blood transfusion must be

correctly identified

throughout the transfusion process from taking the blood sample to the final bedside check to ensure they receive the right blood.

Patients can also form antibodies to many other red cell antigens
The blood selected for transfusion should be

negative

for the red cell antigen(s) to which

antibodies have formed.

All patients receiving blood transfusion should also be monitored for other reactions such as

fever or **allergic reactions.**

Close communication is essential

between clinical teams, the hospital transfusion laboratory and the blood services for safely managing patients with SCD needing transfusion.

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information for patients and relatives

Tell any Doctor or Nurse caring for you that you have Sickle Cell Disease

and they need to let the Transfusion Lab know that in advance, if they are considering giving you a transfusion.

If you have any cards or documents about your special requirements then

show them to the Doctor or Nurse caring for you

and ask them to inform the Transfusion Lab

If you have red cell antibodies then tell the Doctor or Nurse caring for you and ask them to

inform the Transfusion Lab

This will help minimise any delay in finding the right blood for you.

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information for hospitals, doctors and nurses

Always let Transfusion Labs know

that the patient has SCD as they have additional special requirements for blood (eg: blood that is negative for certain antigens, HbS negative etc).

Always use the correct patient ID

including their full name, date of birth AND hospital number / NHS number, so that we ensure that the right patient gets the right blood

SCD patients are particularly prone to

developing red cell antibodies,

with the risk of delayed haemolytic transfusion reactions (renal failure & jaundice), with in some cases, difficulty in sourcing compatible blood.

The development of antibodies may jeopardise their future transfusion programme.

Matching of blood for full Rh (C, c, D, E, e) and K,

reduces the risk of forming these antibodies. Labs can only match if they are informed that the patient has SCD.

If possible,

find out which hospital normally undertakes their care

for SCD and let the Transfusion Lab know (eg: state on Transfusion request form).

Indications for transfusion in SCD patients are increasing

so giving the right matched blood is important, particularly if the patient is not known to the current hospital.

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some improvements in blood provision

Proactive genotyping enables

better selection of blood in emergencies,

when patients have auto-antibodies or a positive DAT which could mask underlying alloantibodies. By matching blood for antigens which patients lack, the risk of haemolysis would be reduced.

Identification of patients with Rh variants

+/- corresponding allo-antibodies, and whether clinically significant haemolysis has occurred, could enable better matched blood provision for patients, or if unavailable, then advice on IVIg cover for transfusion may reduce haemolysis.

Genotyping can be done even when patients have been

recently transfused

(whereas phenotyping cannot).

ADVANTAGES FOR PATIENTS AND THE BLOOD SERVICE

NHS Blood and Transplant (NHSBT) launched a project this year, to

red cell genotype all patients with Sickle Cell Disease, by June 2016.

Red cell genotyping at the International Blood Group Reference Laboratory (IBGRL), Bristol covers Rh variants as well as genes for the usual antigens covered by extended phenotyping (Full Rh, Kell, Fy, Jk, MNSs).

Patients with Rh variants (usually SCD patients) can be clinically assessed proactively to establish whether they have corresponding allo-antibodies +/- haemolysis following transfusion with unmatched blood, and a plan for

the most appropriate blood for transfusion

in future can be made with reference to the relevant expertise & literature, before the need arises.

Proactive genotyping of all SCD patients will enable NHSBT to better

predict the demand

for blood, particularly for “rare” blood due to patients having multiple antibodies or antibodies to high frequency antigens (eg: anti-U).
NHSBT could better plan to meet patient needs through defined plans for extended genotyping or phenotyping of more donors for “rare” blood and recruitment of relevant donors to address unmet demand.

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