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.
SICKLE CELL AWARENESS
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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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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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.
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).

The development of antibodies may jeopardise their future transfusion programme.

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.

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.
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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