**Blood transfusion in patients with Sickle Cell Disease**

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| **Profile** | |
| **Version:** | *V3.1* |
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| **Executive/Divisional sponsor:** | *Dr Lisa Pickering, Divisional Chair (v3.0 2019)* |
| **Applies to:** | *All staff involved in the care of patients with Sickle Cell Disease* |
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| **Approval** | |
| **Approval person/Committee:** | *MedCard Divisional Governance Board* |
| **Date:** | *14th February 2019* |
| **Approval person/Committee:** | *Hospital Transfusion Committee* |
| **Date:** | *March 2020* |

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**Policy Gateway**

Please complete the checklist and tables below to provide assurance around the policy review process.

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| ☒ I have involved everyone who should be consulted about this policy/guidance  ☒ I have identified the target audience for this policy/guidance  ☒ I have completed the correct template fully and properly  ☒ I have identified the correct approval route for this policy/guidance  ☒ I have saved a word version of this policy/guidance for future reviews and reference |

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| Please set out what makes you an appropriate person to conduct this review: |
| Consultant for Adult Haemoglobinopathy service |

|  |
| --- |
| Please set out the legislation, guidance and best practice you consulted for this review: |
| * West Midlands Review Service – Quality Standards: Health Services for People with Haemoglobin Disorders (2018/19) * Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK 2018 (Sickle Cell Society) <https://www.sicklecellsociety.org/wp-content/uploads/2018/04/Web-version-FINAL-SCS-Standards-GSM-6.4.18.pdf> * British Society Haematology : guidelines Red Cell Transfusion in Sickle Cell Disease Part I and Part II (7.11.17 and 18.11.18) <https://b-s-h.org.uk/guidelines/guidelines/red-cell-transfusion-in-sickle-cell-disease-part-l/> |

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| --- |
| Please identify the key people you involved in reviewing this policy why, and when: |
| * Kelly Feane – Lead Transfusion Practitioner |

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| Summarise the key changes you have made and why: |
| Updates in line with the UK standards of care for SCD  Updated contact details  V3.1 February 2020 – Formatting update predominantly |

**Executive Summary**

Transfusion in patients with Sickle Cell Disease (SCD) and thalassaemia is increasing rapidly across the UK but with variability in indications and lack of robust evidence in many cases. Guidance is now issued by both the British Standards of Haematology (2016) and from the Standards of Clinical Care for adult patients with SCD in the UK. These guidelines describe the indications, management and process (including safety and selection) for blood transfusion in SCD and thalassaemia.

## Introduction

Transfusion in patients with Sickle Cell Disease (SCD) and thalassaemia is increasing rapidly across the UK but with variability in indications and lack of robust evidence in many cases. Guidance is now issued by both the British Standards of Haematology (2016) and from the Standards of Clinical Care for adult patients with SCD in the UK. These guidelines describe the indications, management and process (including safety and selection) for blood transfusion in SCD and thalassaemia.

These guidelines should be used in conjunction with the St George’s University Hospitals NHS Blood Transfusion Policy <http://stg1wordpress01/wordpress/wp-content/uploads/2016/06/Blood-Transfusion-Adults.pdf>

## *Status and Purpose*

*This document is part of the Haematology Department’s guidelines on the management of patients with SCD and is applicable to all staff involved in the care of these patients.*

## *Definitions*

*Sickle Cell Disease – inherited lifelong condition due to abnormal haemoglobin variant.*

## *Scope*

*This guideline is relevant to the care of patients with SCD requiring elective and emergency surgery at St.George’s.*

## *Roles and Responsibilities*

### *5.1 Haemoglobinopathy team (Consultant haematologists, Clinical Nurse Specialist and Clinical Health Psychologist) – Responsible for the care of these patients, developing and updating guidelines to be reflective of good practice and to deliver the training to ensure good safe care.*

***5.2 Medical staff involved in the care of patients with SCD.*** *Responsible with the oversight of the haemoglobinopathy team to deliver the care to these patients in line with guidelines where possible.*

***5.3 Nursing staff and allied health professionals involved in the care of patients with SCD on wards, day unit and other areas of St George’s*** *responsible with the oversight of the haemoglobinopathy team to deliver the care of these patients in line with guidelines where possible.*

1. **Contents** 
   1. summary of transfusion in SCD
   2. Indications for top up transfusions in SCD
   3. Indications for elective exchange transfusions in SCD
   4. Indications for emergency exchanges in SCD
   5. Safety and Monitoring of patients with SCD on long term transfusions
   6. Patients who refuse Blood Transfusion
   7. Patients who are ‘difficult to transfuse’

**6.1 Summary of Transfusion in SCD**

In patients with **sickle cell disease** (SCD) the transfusion of packed red cells has 2 goals:

1. To increase tissue oxygenation by correcting anaemia
2. To prevent or reduce the complications of sickling by reducing the HbS% content of the blood

“Top up” transfusions are generally required to achieve the first aim. Exchange blood transfusions usually achieve the second goal.

All **unplanned transfusions** for patients with sickle cell disease must be discussed with the haematology team caring for the patient. In patients with sickle cell disease, mild anaemia does not usually require correction by blood transfusion

The **blood transfusion laboratory** must be made aware that the recipient patient has sickle cell disease or thalassaemia so that extended phenotyping can be done and appropriately crossmatched and sickle negative blood can be provided. These patients are not suitable for rapid cross match.

All patients with a haemoglobinopathy should, at their first engagement with the hospital, have a sample sent to the blood transfusion laboratory for a full, extended red cell antigen phenotype (and genotype where possible) as well as the standard ABO and Rh blood group.

**Guidelines on red cell transfusion in sickle cell disease. Part I: principles and laboratory aspects (BSH) – Summary of Key Recommendations (November 2016)**

* The decision to top up or exchange transfuse an adult or paediatric patient with sickle cell disease (SCD) needs the input of a clinician with appropriate experience. Specialist advice should be obtained for the management of patients with complex transfusion requirements
* Transfusion in SCD requires careful consideration of both the haemoglobin concentration (Hb) and/or percentage of sickle haemoglobin (%HbS) in order to ensure maximal oxygen delivery to tissues without increasing overall blood viscosity to detrimental levels
* A transfusion history should be obtained in all SCD patients requiring transfusion, whether elective or emergency. Close communication is essential between clinical and laboratory teams so that appropriate blood is given
* Individuals with SCD are high‐risk surgical patients. Close liaison between all clinical teams is essential with preoperative optimisation and appropriate postoperative care, whether transfused or not
* Virology testing [hepatitis B, hepatitis C and human immunodeficiency virus (HIV)] should be undertaken at presentation and hepatitis B vaccination should be given to all patients with SCD, irrespective of previous or prospective planned transfusions. SCD patients on regular transfusions should be screened annually for hepatitis B, hepatitis C and HIV
* The choice of transfusion method, i.e., simple (top up) or exchange, should be based on clinical judgement of individual cases, taking into account the indication for transfusion, the need to avoid hyperviscosity and minimise alloimmunisation, maintenance of iron balance, venous access issues and available resources
* All hospitals that are likely to admit SCD patients should have staff trained in manual exchange procedures and clearly identified manual exchange procedures, as this can be lifesaving in emergency situations
* Large referral centres managing patients with SCD should have facilities and trained staff for automated exchange transfusion
* If transfusion is needed, patients with SCD should be given ABO‐compatible, extended Rh‐ and Kell‐matched units. If there are clinically significant red cell antibodies (current or historical) then the red cells selected should be negative for the corresponding antigens
* Patients with SCD must also have extended red blood cell (RBC) antigen typing performed, which may assist with further serological testing and selection of red cell units if there are haemolytic reactions and complex transfusion requirements
* Blood provided for SCD patients should be HbS negative and, where possible, should be <10 days old for simple transfusion and <7 days old for exchange transfusion but older blood may be given if the presence of red cell antibodies makes the provision of blood difficult
* All patients with SCD should carry a transfusion card indicating that they have ‘special requirement’ and, in particular, giving information of any alloantibody.
* Patients with multiple red cell alloantibodies or antibodies to rare antigens need a clear agreed plan given that blood may be difficult to source in the elective or emergency setting. Close liaison between all clinical teams, the hospital transfusion laboratory and the national blood service is essential to ensure appropriate provision of blood.
* All clinicians managing patients with SCD should be aware of the risk of haemolytic transfusion reactions to ensure prompt recognition and management. Close liaison is needed with haemoglobinopathy specialists and blood services for investigation and management.
* Any adverse events or reactions related to transfusion should be appropriately investigated and reported to local risk management systems and to UK Haemovigilance Schemes

**6.2 Indications for Top Up Transfusions**

SICKLE CELL DISEASE

* Symptomatic anaemia (eg haemolysis, sequestration, acute parvovirus infection)
* Consider if Hb <60g/L or if fall from baseline is greater than 20g/L
* Before surgery in some cases in discussion with the sickle cell team and as per their preoperative surgical plan

In patients with SCD who have an Hb >90g/L, top up transfusions should be avoided in patients to avoid the risk of hyperviscosity. The target post-transfusion Hb should usually be around 90-100g/L or the patient’s own baseline Hb.

Venous access is usually peripheral with the aid of ultrasound guidance and should be achieved within 3 attempts (though if more than once is required regularly this should be escalated). Wherever possible ports and indwelling central lines should be avoided in patients with TDT due to increased risk of line thrombosis and infection

**6.3 Indications for Elective Exchange Transfusion in SCD**

* Secondary stroke prevention
* Primary stroke prevention (ongoing transfusion programme from childhood)
* Before elective surgery after discussion with the haematology team

*Consider also in*

* Pregnancy
* Recurrent pain or chest crises not responding to hydroxycarbamide
* Pulmonary hypertension
* Refractory leg ulcers
* Refractory sickle nephropathy / post renal transplant
* Persistent priapisms

All patients being considered for elective exchange transfusions should be discussed at MDT

**6.4 Indications for Emergency Exchange Transfusions**

If Hb <60g/L then a top-up transfusion may be considered initially

In most cases, the target Hct is 30-33% and HbS% < 30%

* Acute stroke
* Acute chest syndrome
* Severe sepsis and acute multi-organ failure
* Progressive intrahepatic cholestasis/Right upper quadrant syndrome
* Hepatic sequestration
* Fulminant priapism not responding to urological intervention

At SGH emergency exchange transfusions are usually automated. The decision to perform an exchange transfusion should be made by the haematology registrar in liaison with the haematology consultant for haemoglobinopathies or the on call haematology team out of hours.

The haematology SpR will contact the on call apheresis team, organise venous access and liaise with the blood transfusion laboratory.

Any patient requiring emergency exchange transfusion will need to be on an HDU bed as a minimum so discussion with critical care is essential. This must happen before agreeing to transfer a patient from another site.

The post transfusion target Hct should be no higher than 33% and HbS% should be <30%.

If it is not possible to perform an automated exchange, then a manual exchange can be done following separate guidelines on manual exchange.

**6.5 Safety and Monitoring of patients with SCD on long term transfusions**

* Blood safety
  + Hepatitis B vaccination and titre checks annually
  + Annual HIV Ag and Ab test
  + ABO‐compatible, extended Rh‐ and Kell‐matched units. If there are clinically significant red cell antibodies (current or historical) then the red cells selected should be negative for the corresponding antigens
  + Less than 10 days old if possible.
* Patient Monitoring

At SGH there are quarterly documented meetings with apheresis nursing staff and medical staff to discuss patients who are on regular transfusion programmes:

* Any predicted issues
* Any reported issues
* Venous access concerns
* Acceptable Hct and S% levels in patients with SCD
* Acceptable pre-transfusion Hb levels in thalassaemia patients
* Review of chelation
* Review of clinic attendance
* Discussion of transfusion and patient associated audits
* Referral of new patients to an apheresis programme
* Referral of new patients to a top up transfusion programme
* Transition of patients from paediatrics to adult care
* Patients stopping a transfusion programme
* Any emergency procedures in the preceeding time frame.

SCD Patients on long term transfusions should be having automated red cell exchanges unless contraindicated

Venous access is usually via ultrasound guided peripheral access – if hard to find after 3 attempts then discussion about central access with the patient should occur

**6.6 Patients who refuse Blood Transfusion**

Patients who refuse blood transfusion for religious or other personal reasons will receive optimum care with wishes respected

The Trust policy on the management of patients who refuse blood should be followed (and can be found on the Red Cell Disorder Unit website or at St George’s University Hospitals Policy Hub) and the trust documentation used.

**For patients with a haemoglobinopathy disorder (sickle cell anaemia/thalassaemia), their blood refuser status and paperwork should be reviewed on an annual basis (where possible), preferably with the red cell haematology consultant and a member of the transfusion team.**

* 1. **Patients who are ‘difficult to transfuse’**

**Patients with multiple red cell alloantibodies or antibodies to rare antigens need a clear agreed plan given that blood may be difficult to source in the elective or emergency setting. Close liaison between all clinical teams, the hospital transfusion laboratory and the national blood service is essential to ensure appropriate provision of blood**

*Patients with a history of hyperhaemolysis are also considered ‘difficult to transfuse’ but this is not due to the availability of appropriate blood units but rather the risk of transfusing any red cells. For these patients please follow the guideline on the management of hyperhaemolysis.*

Multiple antibody combinations or antibodies to rare antigens not seen in Caucasian donors (e.g. Fya‐b‐) can cause difficulty in sourcing blood. The development of anti‐U and anti‐Jsb can be particularly. Alloimmunisation can lead to delays in securing compatible units, as well as increased potential for delayed haemolytic transfusion reactions.

Careful planning and communication between all teams is essential; this includes liaising with the Blood Service for sourcing rare blood from donors and providing frozen units where necessary.

British Society Haematology Guidelines (2016) state that

* The principal aims are to provide compatible blood in a timely manner and to minimise the risks from transfusion.
* Patients who are difficult to transfuse should have a written plan detailing: the transfusion requirements, a contingency plan for emergency transfusion, the contact details of the patient's sickle/haematology consultant and a recommendation to involve a National Blood Service reference laboratory.
* In England, NHSBT provides hospital transfusion laboratories access to **Sp**ecialist Services Electronic Reporting System, delivered via Sunquest **ICE** (Sp‐ICE) for review of patient's serological records (<http://hospital.blood.co.uk/diagnostic-services/sp-ice-browser/www.hospital.blood.co.uk>).
* For long‐term management, hydroxycarbamide can reduce the need for transfusions and should be considered (Charache *et al*, [1995](https://onlinelibrary.wiley.com/doi/full/10.1111/bjh.14346#bjh14346-bib-0019)).

At St George’s we undertake annual audits of alloantibody formation and existence. Patients with a complex antibody status are identified and discussed at a quarterly haemoglobinopathy transfusion meeting.

Patients should have need for transfusion reviewed by consultant in haemoglobinopathy and other disease intervensions considered (eg hydroxycarbamide, novel therapies)

In particular patients are identified who would fulfil the following criteria

* History of hyperhaemolysis
* at least 3 alloantibodies
* anti-U
* anti-Fya and anti Fyb together (antiFy3)
* anti-Jsb
* presence of an antibody to which antigen negativity occurs in less than 5% of the current donor population in England; e.g. Kp(b), k, Lu(b), (+others?)
* U negative antigen

## 

## Dissemination and implementation

### 7.1 Dissemination:

Guidelines will be available on the departmental intranet page and available in paper form in the junior doctor office in haematology.

### 7.2. Implementation

Guidelines will be promoted by the haemoglobinopathy team.

## Consequences of Breaching the Policy

## Failing to follow this policy could lead to action under the Trust’s disciplinary policy.

## Monitoring compliance

The table below outlines the process for monitoring compliance with this document.

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| **Monitoring compliance and effectiveness table** | | | | | |
| **Element/ Activity being monitored** | **Lead/role** | **Methodology to be used for monitoring** | **Frequency of monitoring and Reporting arrangements** | **Acting on recommendations and Leads** | **Change in practice and lessons to be shared** |
| *WMQRS peer review quality standards* | *Consultant haematologist* | *As required (every 2-3 year)* | *The lead or committee is expected to read and*  *interrogate the report to*  *identify deficiencies in the system and act upon them. Consider stating this responsibility in committee terms of reference.* | *Required actions will be identified and completed in a specified timeframe.*  *Consider stating this responsibility in committee terms of reference.*  *These will be discussed at Divisional governance board* | *Required changes to practice will be identified and actioned within a specific timeframe. A lead member of the team will be identified to take each change forward where appropriate. Lessons will be shared with all the relevant stakeholders.* |

## Associated documentation

Incentive Spirometry guideline for adults admitted with sickle cell crisis at risk of acute chest syndrome

## References

* West Midlands Review Service – Quality Standards: Health Services for People with Haemoglobin Disorders (2018/19)
* Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK 2018 (Sickle Cell Society) <https://www.sicklecellsociety.org/wp-content/uploads/2018/04/Web-version-FINAL-SCS-Standards-GSM-6.4.18.pdf>
* British Society Haematology : guidelines Red Cell Transfusion in Sickle Cell Disease Part I and Part II (7.11.17 and 18.11.18) <https://b-s-h.org.uk/guidelines/guidelines/red-cell-transfusion-in-sickle-cell-disease-part-l/>