Peri-operative Management of Sickle Cell Disease

Profile	
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Executive/Divisional sponsor:	*Dr Lisa Pickering, Divisional Chair*
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Approval person/Committee:	*MedCard Divisional Governance Board*
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Policy Gateway

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

☒ 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

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-I/

Please identify the key people you involved in reviewing this policy why, and when:

Summarise the key changes you have made and why:
Updates in line with the UK standards of care for SCD Updated contact details

Executive Summary

Patients with sickle cell anaemia undergoing surgery are at an increased risk of complications. This guideline outlines the peri-operative management of patients with sickle cell disease undergoing elective and emergency surgery.

1. Introduction

Elective and emergency surgery is associated with a higher risk of peri-operative complications in patients with Sickle Cell Disease (SCD), including an increased risk in peri-operative mortality, vaso-oclusive crisis, acute chest syndrome and infections. Careful pre-operative planning and assessment is vital in order to minimise the risk of these complications and requires close collaboration between haematologists, surgeons and anaesthetists.

2. 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.

3. Definitions

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

4. Scope

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

5. 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.

6.0 Content

6.1 Pre-operative planning
6.2 Intra-operative care
6.3 Post-operative management
6.4 Contact details

6.1 Pre-operative planning

Patients should be managed jointly by **haematology, surgery and anaesthetics**. For **elective** surgery, the haematology team should be informed as early as possible and **at least 3 weeks before** the date of surgery, so that a blood transfusion (top-up or exchange) can be arranged, if indicated.

Patients will be seen in the pre-operative assessment clinic and their sickle cell status will be confirmed if there are uncertainties, as well as baseline organ function assessment undertaken, such as the organisation of echocardiograms.

Each patient should have an **individualised management plan**, which will depend on the risk of the procedure and the patient's risk factors. This should be communicated to the surgical and anaesthetic teams and filed in the patient's medical records.

Transfusion:

- Transfusion in those with HbSS undergoing moderate and high risk surgery has been shown to reduce the rate of post-operative sickle cell complications. Transfusion may be indicated to decrease the risks of surgery depending on the phenotype and genotype of the patient and the type of surgery required.

- In patients who are anaemic (Hb <80g/L) a simple top up transfusion may suffice to increase the haematocrit and oxygen carrying capacity and offer a moderate dilution in HbS percentage whilst in patients with a Hb >80g/L, those undergoing major surgery (cardiovascular or brain surgery) and those with a severe phenotype (including recurrent ACS, silent strokes or previous problems following an anaesthetic) an automated exchange transfusion can be arranged in the week before the surgery to provide a more robust reduction in the HbS%.

- Rarely, alternatives such as hydroxycarbamide and erythropoietin may be indicated, for example, if the patient has a history of hyperhaemolysis (a life-threatening haemolytic transfusion reaction) or is a blood-refuser.

- Patients should have **repeat group and save samples on admission** being aware that these patients are NOT suitable for rapid cross match.

Patients are usually not suitable for day case surgery and should be **admitted to a ward**, even following minor surgery for observation overnight post-operatively, unless the surgery is performed under local anaesthetic.

If the patient is to be nil by mouth, they should be admitted the evening before surgery and prescribed **IV fluids**. If an **HDU/ICU** bed is required post-operatively this should be organised in advance by the surgical team.

For elective surgery, patients should be placed at the **beginning of the theatre list** to reduce the risk of cancellation, particularly if they have been transfused pre-operatively.

Following **emergency surgery**, the majority of patients should be admitted to **HDU/ICU**. These patients should be **discussed urgently with the haematology SpR/consultant**. Pre-operative transfusion may not be possible but may be indicated post-operatively.

6.2 Intra-operative care

Hydration – volume status should be monitored closely throughout the procedure with avoidance of hypovolaemia.

Oxygenation – Avoidance of hypoxia is important to prevent sickling and tissue ischaemia. Ensure careful oxygenation throughout the procedure and until fully awake.

Temperature regulation – the patient should be kept normothermic throughout the procedure. A Bair Hugger and warmed IV fluids should be considered during long procedures.

Intra-operative cell salvage is **contraindicated** in patients with sickle cell disease.

6.3 Post-operative management

Admission to a **HDU/ITU** bed should be considered post-operatively.

IV fluids should be continued until the patient's oral intake is adequate.

Pulse oximetry monitoring should be continued for at least 24 hours post surgery and oxygen saturations should be maintained at >95%. Incentive spirometry should also be used (see incentive spirometry guidelines for adults admitted with sickle cell crisis at risk of acute chest syndrome). Prophylactic CPAP should be considered following major abdominal and thoracic surgery in high risk patients.

Analgesia – patients may be relatively insensitive to opiate analgesia and require higher doses in the post-operative period. It is therefore important to take into account previous history of opiate exposure when prescribing analgesia. Advice should be sought from the acute pain team if needed.

VTE prophylaxis – thromboprophylaxis (eg. dalteparin 5000 IU od) should be prescribed unless contraindicated. Follow guidelines for extended prophylaxis depending on the type of surgery.

Infection management – patients with sickle cell disease are at an increased risk of infection post-operatively. If the patients becomes febrile, blood cultures should be taken and intravenous antibiotics commenced as per the Trust guidelines.

Emergency exchange transfusion should be considered for all patients with sickle cell disease who have undergone major emergency surgery without prior transfusion.

6.4 Hospital Contact Details

Dr Elizabeth Rhodes	Haematology Consultant	ext 0885
Dr Julia Sikorska	Haematology Consultant Haematology Registrar	ext 0885 bleep: 7080
Carol Rose	Sickle Cell CNS	mobile: 07500835735

Out of hours

Haematology Registrar	via switchboard
Haematology Consultant	via switchboard

7. 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.

8. Consequences of Breaching the Policy

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

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

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.*

10. Associated documentation

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

11. 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-I/