1. Introduction and Who Guideline applies to

This document aims to provide guidelines for peri-operative plans of patients with Sickle Cell Disease (SCD). It applies to all staff who are involved with patients with sickle cell disease undergoing surgery, both in the elective and emergency setting. This guideline is for all patients managed across the East Midlands region and specific information for individual trusts can be found in the appendix.

2. Guideline Standards and Procedures

Patients with Sickle Cell Disease (Homozygous sickle cell anaemia, HbSC disease, HbS beta thalassaemia, HbSD Punjab disease, HbSO Arab) have an increased risk of sickle crisis, thrombotic complications and infection in the peri-operative period. The risk to individual patients will vary, depending on factors including the sickle cell genotype, patient age, evidence of end organ damage, type of surgery etc.

It is therefore important to plan surgery carefully and in advance, to reduce these risks as far as possible.

Planning the procedure

- Where a patient with SCD requires surgery, either planned or emergency, consultation with the responsible haematologist must be documented, in order to review specific requirements and measures to be taken to avoid complications in each case.
- In emergency scenarios, the on call haematology registrar can be contacted via switchboard. If required, cases can be escalated to the regional Specialist Haemoglobinopathy teams based at Nottingham University Hospitals or University Hospitals of Leicester.
- If emergency surgery is urgent and potentially lifesaving, it should be performed as necessary and transfusion (top up or exchange) can be performed intra- or post-operatively.
- In elective cases, the consultant haematologist will review the case and refer to the regional MDT for discussion (if possible given time constraints). This will include a review regarding the need for blood transfusion.

- A peri-operative plan will be produced (see appendix 2 for example template). This should include details of baseline blood levels, blood group, antibodies, viral status, vaccination status, medication, end organ involvement, medical history, medication. Surgical details should include procedure, surgeon and date of procedure (if known)
- In some cases an anaesthetic review prior to admission may be required particularly if the patient has end-organ damage. CXR, Echocardiography, ECG and pain management will need to be reviewed.
- Specific requirements for respiratory support and physiotherapy will need to be identified.
- Need for post-operative HDU admission will be a joint decision between the surgeon, anaesthetist and responsible haematologist.
- A decision to offer pre-operative transfusion will be made in line with the national sickle cell standards. This may be a top-up or exchange transfusion depending on the patient's baseline haemoglobin and sickle cell genotype (see appendix 3).
- Where transfusion/exchange preoperatively is planned, this must be prescribed and organised in good time. Transfused blood must given in line with the trust red cell transfusion in sickle cell disease guideline (Minimum requirements: ABO, Rh, Kell matched and HbS negative).

Pre-operative checklist

A pre-operative check list will aid delivery of appropriate measures to prevent peri-operative complications.

For elective procedures, on the day of surgery:

- Inform local haemoglobinopathy team (see appendix 1). If unable to contact, inform the haematology on call team via local contact mechanism.
- Prescribe usual medication (continue hydroxycarbamide unless neutropenic, until nil by mouth).
- Review usual analgesia.
- Document any allergies.
- Ensure baseline assessment of cardiac and respiratory systems is documented notes. Include review of the following:

CXR ECG Echo O2 sats (on air)

- If not done at pre-assessment clinic, perform baseline blood tests (FBC,U&E,LFT,CRP) and urine dipstick. Rule out evidence of infection prior to surgery.
- Anaesthetic review (prior to admission if known end organ damage)
- Plan perioperative analgesia
- Physiotherapy review (chest physio/plan CPAP/ assisted spirometry, review mobility)
- Haematology review (Registrar or consultant)

Monitoring and supportive measures

Observations

- Review pulse, blood pressure, temperature, respiratory rate, oxygen saturation and pain score throughout the peri-operative period.
- Escalation should be performed in line with trust protocol. Haematology should be informed of any deterioration.

Hydration

- As soon as patient is nil by mouth, give IV fluids maintaining input of 3 litres per day.
- Keep accurate fluid balance chart.
- Continue IV fluids until patient is mobile and drinking freely.

Warmth

To avoid precipitation of a sickling episode:

- Keep patient warm at all times (particularly in transit, anaesthetic room, theatre and recovery)
- Give warmed IV fluids if necessary.

Oxygenation

- Record baseline blood gases if necessary.
- Measure O2 saturation and respiratory rate at least 4-hourly for at least 48 hours post operatively and while receiving opiate analgesia. Sats <95% on air should be escalated to the haematology team.
- Regular physiotherapy review.

Analgesia

- Ensure regular and adequate pain relief, taking regular analgesia into account.
- Use pain scale on EWS chart 4hourly at least, until pain free and fully mobile. Many patients will be opiate naïve and standard post-operative analgesia is appropriate.
- Avoid prolonged use of NSAIDs and prescribe laxatives, especially if opiates used.

Risk of Infection

- Patients are asplenic and at increased risk of infection. Any signs of sepsis should be managed in line with the local sepsis policy.
- Use standard surgical prophylaxis and continue penicillin (or erythromycin) prophylaxis until nil by mouth. This should be continued post-operatively unless treatment antibiotics are prescribed.

Risk of thrombosis

- Patients are at higher risk of thrombosis and all should receive LMWH prophylaxis unless contraindicated.
- Anti-embolic stockings can be offered, but can cause a tourniquet effect in some patients.
- In high VTE risk surgery (such as joint replacement surgery), consider use of flowtron boots until patient is mobile (in line with local thromboprophylaxis guideline).
- Encourage hydration and early mobilisation.

Prior to discharge

- Inform the haematology team on discharge to ensure follow up.
- Post operative blood tests should be reviewed to look for evidence of infection.
- Ensure regular medication prescribed with adequate analgesia on discharge.

Appendix 1: UHL specific information

How to contact Haemoglobinopathy team for advice:

Advice from the haematology team can be sought via any of the following:

Haemoglobinopathy nurse specialist (Mon-Fri) 07950891490

Haemoglobinopathy Team office (Mon-Fri) 0116 258 6081 (Extension 16081)

Haematology registrar (24/7) – Available via switchboard

Elective Procedures

Where possible, all cases should be referred to the EMSTN MDT for discussion before surgery is scheduled.

Please contact uho-tr.emstnhcc@nhs.net for advice on how to arrange this

The decision to transfuse pre-op will be made by the haematology team and communicated to the surgical team.

If required, transfusion will be booked and arranged by the haematology team and be reflected on the pre-operative plan

Emergency Procedures

ANY patient admitted sickle cell disease should be discussed with the haematology team (contact details as above)

If emergency surgery is urgent and potentially lifesaving, it should be performed as necessary and transfusion (top up or exchange) can be performed intra- or post-operatively.

UHL may be contacted from Network sites to facilitate transfer for surgery: Transfer between hospitals should not delay urgent surgery if this is required

Post-operative management

The Haemoglobinopathy team will liaise with surgical teams and arrange post-operative review as necessary

Please avoid triggers for crisis and ensure any pain is managed in line with the UHL policy for acute complications in sickle cell disease

Any plan for discharge should be communicated with the haemoglobinopathy team

Appendix 2: Standard pre-operative plan template

EMSTN East Midlands Sickle Cell and Thalassaemia Network

Private and Confidential

University Hospitals of Leicester NHS — Caring at its best

Pre-operative plan for patients with sickle cell disease undergoing surgery

Personal details		
Patient name:	S number:	DOB:
Primary haemoglobinopathy centre:		
Steady state values: Hb Bilirubin Creatinine Retics Sats on air		

Background	
Known chronic disease complications	Transfusion history **Blood group:** **Known red cell antibodies:**
Current treatment	Other concerns/issues **Echo result:** **Vaccination history:**

Planned Operation:	Date of Procedure	Consultant Surgeon:

Pre-op transfusion: Required Not Required Plan:

Anaesthetic implications: **Analgesia:** *[Include patient's usual analgesia plan]*

Throughout perioperative period, review the following regularly:	
Thromboprophylaxis: Weight appropriate LMWH should be given in line with hospital policy.	
Hydration • As soon as patient is nil by mouth, give iv fluids maintaining input of 3 litres per day • Keep accurate fluid balance chart • Continue IV fluids until patient is mobile and drinking freely	Warmth To avoid precipitation of sickling episode: • Keep patient warm at all times (particularly in transit, anaesthetic room, theatre and recovery). • Give warmed IV fluids if necessary
Oxygen • Measure O2 saturation and respiratory rate at least 4hrly for at least 48 hours post operatively, and every 2 hours whilst receiving opiate analgesia • Regular physiotherapy review and offer incentive spirometry	Infection risk • Continue patients own antibiotic prophylaxis throughout admission, unless treatment dose antibiotics are required. In this case, prophylaxis can he held and restarted when treatment has been completed. • Sickle cell patients are immunosuppressed and if features of sepsis, they should be managed on the sepsis six pathway with early introduction of antibiotics (fever >38, rising CRP/WCC)

Please inform the haemoglobinopathy team on extension 6081 or call CNS on 07950 891490. If out of hours, please call haematology registrar on call via switchboard

Completed by: *Date:*

Appendix 3: Guidelines for pre-operative transfusion

Elective surgery:

If HbSS and HbSB^{0}thalassaemia genotype:

All patients with HbSS and HbSB^{0}thalassaemia should be offered pre-operative transfusion 2-10 days prior to surgery.

Minor or moderate risk operations:

Hb <90g/l – Top up transfusion aiming for pre-op Hb of 100g/l

Hb >90g/l – Partial or full exchange transfusion (to be decided by sickle team)

If patients are already on a regular exchange transfusion programme they should have an exchange timed in the 2-10 days prior to surgery. Some patients with Hb<90g/l and a very severe phenotype will be recommended to have a pre-operative exchange transfusion

High risk operations:

Exchange blood transfusion 2-10 days prior to surgery aiming for pre-op Hb of 100g/l and HbS% of <60%.

For all other genotypes (including HbSC) the evidence for the role of transfusion is less clear. Each case will be reviewed by the haematology team on a case by case basis. In the presence of severe disease phenotype and/or high risk surgery, pre-operative exchange transfusion may be required

Emergency surgery:

Any patient with sickle cell disease requiring emergency surgery must be discussed with haematology on call. The decision to transfuse will depend on genotype, phenotype, required surgery and pre-operative haemoglobin.

If surgery is urgent and potentially lifesaving, it should be performed as necessary and transfusion (top up or exchange) can be performed intra- or post-operatively.

3. Education and Training

None

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Peri-operative plan completion for patients with sickle cell disease undergoing surgery	Network audit	Dr Amy Webster	Every 2 years	EMSTN network

5. Supporting References (maximum of 3)

Guy's and St. Thomas' NHS Trust Clinical guideline for peri-operative management of sickle cell disease in adults.

Standards for the clinical care of adults with sickle cell disease in the UK, 2nd edition 2018

Guideline on the peri-operative management of patients with sickle cell disease: Guideline from the Association of Anaesthetists 2021

6. Key Words

List of words, phrases that may be used by staff searching for the Guidelines on PAGL. If none – sickle cell, surgery

CONTACT AND REVIEW DETAILS	
Guideline Lead (Name and Title) Dr Amy Webster, Consultant Haematologist	**Executive Lead**
Details of Changes made during review: Change in staffing contacts to reflect new working practice	