

STSTN GUIDELINE FOR THE PERI-OPERATIVE MANAGEMENT OF SICKLE CELL DISEASE IN ADULTS

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Elective and emergency surgery is associated with significant increased risk for patients with sickle cell disease. The following guideline addresses the risks and pre-emptive management of sickle cell patients undergoing surgery.

The following key areas need to be addressed:

- **Pre-operative planning**

Patients should be managed jointly between haematology, surgery and anaesthetics, with close peri-operative liaison. The Haematology team should be informed as far in advance of elective surgery as possible.

- **Emergency Surgery**

For emergency surgery, there will not always be time for top-up or exchange transfuse pre-operatively. Patients will often need to go to critical care. *Emergency exchange transfusion should be considered for all sickle cell disease patients on critical care wards or who have undergone major emergency surgery without prior exchange transfusion.*

- **Elective Surgery**

Most elective surgery should be carried out in a specialist centre.

Patients who do not attend a specialist centre should be referred to a specialist haemoglobinopathy team centre, as well as to the relevant surgical team in the specialist centre, processes which will take at least eight weeks prior to the elective surgery.

A clear and expeditious referral pathway should be in place for patients being referred from local units to specialist haemoglobinopathy centres for their surgery.

Early referral to the surgical and haematology teams in a specialist haemoglobinopathy centre (SHC) is recommended. This will allow review by both specialties in the SHC in advance of the proposed surgery and ensure that a perioperative transfusion plan, where required, can be agreed and arranged.

As soon as the surgical team make a decision to proceed with an operation the Sickle cell team should be informed to review the patient and agree a pre-op transfusion plan. The pre-op assessment team should also inform the sickle cell team once the patient has been assessed to confirm the Sickle Cell pre-operative transfusion plan.

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A written individual management plan should be in place prior to surgery, which takes into account the patient's risk factors and the type of procedure planned. This plan should be accessible to or circulated to all relevant individuals prior to an elective surgical procedure.

The patient should be informed and involved in their management plan, and should be provided with a copy of their individual sickle peri-operative management plan.

Patients may have a history of chronic pain and be relatively insensitive to opiate analgesia. Their previous analgesia history and care plan should be taken into account when prescribing in the post-operative period. Consider referral to the acute pain team for patients who are taking regular opioids at home or where difficulties with pain relief are anticipated.

In general, day case surgery is usually not suitable for this patient group, except for eye, dental surgery, and surgery under local anaesthetic. Patients will usually require admission overnight post-operatively, for observation in most cases. If a high dependency or Intensive care unit admission is anticipated, necessary arrangements should be made in advance by the surgical team.

Sickle patients should be placed near the beginning of the theatre list to minimise time fasting and reduce likelihood of cancellation if possible, particularly when patients have had a pre-operative blood transfusion. It is acknowledged that this will not be possible in certain circumstances such as patients with a history of MRSA or VRE infection or colonisation.

Blood Transfusion Guidance

- **Transfusion prior to elective surgery**
- Patients undergoing major surgery and those with a history of previous sickle related complications are at higher risk in the peri-operative period. Transfusion decisions should be made after considering patient risk factors and the nature of the planned surgery.
- **1. Low Risk**
 - Even low risk patients undergoing minor procedures under general anaesthetic with a low risk of complications require discussion with Haematology. Top up transfusion should be considered, aiming for pre-operative Hb of 100 g/l for HbSS and HbSβ⁰ patients.
- **2. Intermediate Risk**
 - Patients with a previous history of chest crisis (or other co-morbidity) undergoing low risk surgery; or patients undergoing Intermediate risk surgery (e.g. tonsillectomy, splenectomy, cholecystectomy or joint surgery other than total hip replacement) with no additional patient risk factors.
 - Top-up transfusion aiming for Hb of 100 g/l (irrespective of HbS %)

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- **3. High Risk**
 - High-risk surgical procedures (e.g. cardiothoracic, major upper abdominal surgery, neurosurgery, total hip replacement) or previous serious sickle related complications (e.g. severe chest crisis, CVA, ITU admissions), irrespective of surgical risk.
 - Red cell exchange transfusion (preferably automated) aiming for pre-operation HbS % of <10-30%

- In patients with a high baseline Hb >95g/dl, avoid top up transfusion to prevent hyperviscosity. Particular care is needed in patients with HbSC disease as they are more likely to develop hyperviscosity related complications with top-up transfusion. Exchange transfusion is likely to be needed for all but low-risk procedures.
- Blood should be sickle negative and matched for Rh and Kell antigens and be negative for any known red cell allo-antibodies, as a minimum.
- These principles are also applicable to patients undergoing emergency surgery if time permits. As a minimum these patients should have suitable cross-matched blood available in case an emergency exchange procedure is needed. Haematology team and Blood Transfusion laboratory should be informed on admission if surgery is likely.

- **Patients may have red cell alloantibodies which can delay issue of appropriate cross matched red cells for transfusion.**

AHA/ACS classification:

High risk (cardiac risk >5%)	Intermediate risk (cardiac risk 1–5%)	Low risk (cardiac risk <1%)
Open aortic surgery	Elective abdominal surgery e.g. laparoscopic cholecystectomy	Dental
Major vascular surgery	Carotid endarterectomy	Ophthalmic
Peripheral vascular	Endovascular aneurysm	Incision and drainage of an abscess
Urgent body cavity	Head and neck surgery	Medical termination of pregnancy
Total hip replacement (THR)	Major Gynaecological - hysterectomy	Minor Gynaecological – e.g. hysteroscopy, dilatation and curettage, ERPC
Major neurosurgery	Major reconstructive surgery	Leg ulcer debridement
	Joint surgery (excluding THR)	Minor orthopaedic procedures e.g. core decompression
	Major urological surgery	Minor urology e.g. cystoscopy
	Elective pulmonary surgery e.g. pneumonectomy	Bronchoscopy/EBUS
	Mastectomy	Breast mass biopsy/Lumpectomy

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	Thyroidectomy/parathyroidectomy	Thyroid biopsy
		ERCP/Endoscopy

Modified from Fleisher LA, Beckman JA, Brown KA, et al. ACC/AHA 2007 Guidelines on perioperative cardiovascular evaluation and care for non-cardiac surgery: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on practice guidelines.

- **Patients who cannot be transfused (patient choice or rare blood type)**

Rarely, treatment with Hydroxycarbamide with or without erythropoietin, under the advice of Haematology, may be considered to improve the haemoglobin pre-operatively usually if:

- Difficulties in transfusion are anticipated e.g. a history of repeated delayed haemolytic transfusion reaction despite steroid and IVIg cover, rare blood type due to red cell antibodies such as anti-U.
- Patients who have had a DHTR despite steroid and IVIg therapy should be referred to the regional MDT pre-operatively and considered for Rituximab pre-treatment or any other available therapies.
- Patient declines the use of blood products (e.g. Jehovah's Witnesses).

For patients who are declining the use of blood products in advance of an operation, a clear advance directive stating clearly which products the patient is declining and any potentially useful product that the patient will accept must be documented and accessible/shared with the anaesthetic, surgical and sickle team.

- **Pre and peri-operative management**

- 1. Pain relief**

The patient's usual opiate requirement should be taken into consideration, when managing post-operative pain. Their individual care/analgesia plan should be reviewed and updated prior to admission for surgery. Referral to the acute pain team may be necessary in patients with a high baseline opiate requirement. Pain management in the post-operative period should be led by the anaesthetic and acute pain teams with input from the sickle teams. Where a pre-operative analgesia plan is agreed this should be instituted. Sickle patients with a history of chronic pain may be relatively opiate resistant and require higher doses for adequate analgesia. There is a high incidence of painful crises in the post-operative period.

- 2. Hydration/fluid balance**

Maintain optimum hydration and start IV fluids when oral fluids are stopped. Consider admission the evening before surgery for overnight hydration in patients who are unable to drink up to two hours before surgery, continue IV fluids until the patient is able to take fluids freely. Careful attention needs to be paid to fluid balance.

- 3. Oxygenation**

Avoidance of hypoxia is vital to prevent sickling and tissue ischemia. Document baseline oxygen saturation (possible pre-existing cardio-respiratory disease) and ensure careful oxygenation from pre-medication, throughout the operation, until fully awake. In the immediate post-operative period patients should be closely monitored, with at least hourly observation for the first 6 hours. Any desaturation to oxygen saturations <94% (depending on usual base-line oxygenation) should be reported to medical staff and corrected with supplementary oxygen. Please escalate to the haematologist and the anaesthetist in the

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immediate peri-op period. Escalate urgently if increasing oxygen requirements to maintain saturations >95% or failure to attain this despite oxygen therapy. Consider use of an incentive spirometer in sickle patients post operatively and encourage early mobilisation with physiotherapy input if appropriate.

4. Temperature Regulation

Hypothermia can trigger peripheral stasis and sickling. Keep the patient normothermic during surgery, using warm air blankets and adjust the ambient temperature in theatre as necessary. Ensure anaesthetic team is aware of these requirements in advance.

5. Prophylactic antibiotics and Infection management

Choice and duration of prophylactic antibiotics will depend on the nature of the surgery undertaken. Additional antibiotic prophylaxis should be detailed in the patient's perioperative management plan. Patients should recommence their routine prophylaxis with Penicillin V 250mg bd (or erythromycin 250mg bd in penicillin allergic individuals) once the surgery specific antibiotic course is completed. Sickle patients are functionally hyposplenic and are at increased risk of perioperative infections, if a patient develops a fever (>38.0°C) ensure blood cultures are taken and start intravenous antibiotics as per local antimicrobial policy (NB hyposplenic and regular penicillin V exposure as prophylaxis). Check cannula and line sites daily for signs of infection and monitor for symptoms such as productive cough, shivering or myalgia.

Elective surgery should not proceed in the presence of active infection as this will increase the risk of serious sickle related complications.

6. Thromboprophylaxis

Sickle cell patients are at increased risk of venous thromboembolism. Thromboprophylaxis should be considered for all procedures, particularly for major surgery or if patient will be immobile for >24 hours post procedure. Pharmaceutical thromboprophylaxis (e.g. enoxaparin 40 mg od) should be considered unless contra-indicated. See adjustments for extremes of weight and abnormal renal function as per Trust LMWH and Unfractionated Heparin Dosing and Administration Guidelines, and follow guidelines for extended thromboprophylaxis as per surgery type.

7. Tourniquets are contraindicated in patients with Sickle Cell disease.

8. Cell Salvage - intra-operative cell salvage is contraindicated in patients with Sickle Cell Disease.

9. Admission general points

- Ensure the Sickle Cell team is made aware of the admission for surgery as soon as possible. In addition to FBC, HbS% or HbS+C% and routine bloods, a sample for Group and Save, antibody screening (and red cell phenotyping only if not performed previously) should be sent on admission.

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- Emergency surgical procedures in patients with sickle cell disease should always be discussed immediately with the duty Haematology ST3+ or Consultant, as a peri-operative or immediately post-operative top up or exchange transfusion is likely to be required.
- Do not transfuse sickle patients without prior discussion with the Haematology/sickle cell team, unless lifesaving.
- Admit on the day before surgery if possible.
- All post-operative sickle cell inpatients should be seen regularly by the Sickle Team.
- If the post-operative sickle cell patient develops new symptoms suggestive of a sickle related complication, such as a painful crisis, chest crisis or CVA, the sickle team should be informed immediately.

Individual Sickle Peri-Operative Management Plan

Personal Details:	
Surname:	Hospital No.:
Forenames:	Sex:
D.O.B.:	NHS No.:
Usual specialist/local specialist haemoglobinopathy centre:	
Haematology Consultant:	
Surgical consultant:	
Clinical Details:	
Sickle Genotype: HbSS / HbSβ ⁰ / HbSC / HbSβ ⁺ / other	
Planned surgery: _____	
Operation date: _____	
Red cell alloantibody: none/red cell alloantibody present (details)_____	
Check with blood bank whether difficulties with blood provision YES/NO	
History of haemolytic transfusion reaction: YES/NO	
History of acute chest syndrome or complication requiring emergency exchange transfusion or ITU admission: YES/NO	
Other medical co-morbidities: _____	
Echocardiogram required: YES/NO	
HDU/ICU admission anticipated? YES/NO If YES, surgical team to book bed	
Blood transfusion:	
For peri-operative transfusion?: Yes/No Automated red cell exchange/Top up (delete)	
Venous access for exchange (delete): transfusion: peripheral / portacath / femoral line	
Date of planned pre-operative transfusion: _____ Location:_____	
Does patient refuse blood products? YES/NO	
Has a blood product refusal proforma been completed? YES/NO	

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Steady State Values:	Hb (g/dl): Platelets:	O ₂ on air (%): BP:	Creatinine: Bilirubin:	ALT: Weight:
Analgesia Care Plan:				
<i>Inpatient:</i> e.g. Morphine Sulphate 10mg-20mg s/c 2-4 hourly or oxycodone 5-10mg s/c 2-4 hourly				
<i>Outpatient:</i> e.g. Co-codamol 30/500 2 tablets qds PRN po Ibuprofen 400mg tds PRN po Paracetamol 1 prn qds PRN po				
Allergies:				
Other Medication:				
e.g. Folic Acid 5mg od, Penicillin V 250mg bd				
Anticoagulant and antiplatelet agents(detail):				
Is patient on warfarin/rivaroxaban/apixaban/dabigatran/ agent/ aspirin/clopidogrel/ other anticoagulant or antiplatelet? (delete as applicable and give details):				
Has an anticoagulation bridging plan been completed? YES/NO				
Omit warfarin for 5 days prior to invasive procedures, including femoral access for red cell exchange				
Date completed:		Completed by:		Signature:

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