

## Clinical Guidance

# Perioperative Management of Sickle Cell Disease in Adults

### Summary

*This trust sees large numbers of patients with sickle cell disease and sickle cell trait and many of these patients undergo surgery. Surgery is a time of high risk for patients with sickle cell disease and is associated with many complications including an increasing risk of painful crisis and acute chest syndrome. It is therefore essential that we have a robust mechanism to identify and manage patients with sickle cell disease to prevent peri-operative complications.*

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## **Introduction:**

There are thought to be more than 10,000 patients with sickle cell disease (SCD) in the UK, the majority live in London. Homozygous sickle cell anaemia (HbSS) is the most common and most severe form of SCD in the UK, accounting for 70% of the patients. Compound heterozygotes for HbS and HbC (HbSC) account for the majority of the remainder. Patients with sickle cell trait (AS - carriers) have symptoms only in very extreme circumstances. This Trust sees large numbers of patients with SCD and sickle cell trait. This protocol aims to provide advice on the basic, minimum standard of care for patients requiring anaesthesia. Sickle cell disease is associated with increased risks of peri-operative complications which can be decreased by multidisciplinary pre-operative care.

Universal newborn screening for SCD was introduced in the UK in 2006, and has been carried out locally for several years, but individuals who are older, or who were born outside the UK may never have been screened.

## **Pre-operative identification of patients:**

1. It is medically prudent to screen the sickle cell status of all non-Northern Europeans prior to surgery if their haemoglobinopathy status is not known. All elective non-Northern European patients are screened for a haemoglobinopathy in the pre-assessment clinic, unless they already have a result on EPR (order 'Sickle Cell and Thalassaemia screen').
2. The laboratory will screen for sickle cell status using the full blood count and film, and HPLC (High Pressure Liquid Chromatography) which characterises the different haemoglobinopathies. HPLC takes about one hour to complete.
3. If the sickle cell status (genotype) is unavailable on the day of the operation, defer the case to the end of the list. Take a blood sample (3ml EDTA (lilac topped tube) or 1ml in a Paediatric EDTA tube) to the Haematology Special laboratory, ex 83421 and request sickle cell testing and a full blood count and film. HPLC will be performed urgently if requested directly and can be available within a few hours i.e by the end of the operation during working hours.
4. If the patient requires immediate anaesthesia during working hours, do as in (3) and treat the patient initially as if they have sickle cell disease. The full sickle cell status using HPLC should be available within a few hours i.e by the end of the operation. In the interim the following interpretation from the full blood count should be applied:

If a patient has a normal haemoglobin concentration and does not have a history of painful crises then Hb SS is unlikely. However a normal haemoglobin may be compatible with HbSC disease. In HbSC disease sickling can occur. In Hb SC splenomegaly is common and target cells are found on the blood film. If there is doubt as to whether a patient has sickle cell trait (heterozygote) or sickle cell disease, please discuss all the Haematology results with the Sickle Cell SpR (Bleep 0248), the on call haematology SpR out of hours or a Haematology Consultant.

5. Out-of-hours HPLC is available at Guy's site and turn round time may be slower due to transport and manpower issues. All patients at risk should be screened for sickle

cell disease with a full blood count and HPLC and if there is doubt about the diagnosis then please discuss all the Haematology results with the On call Haematology SpR or Consultant (contact via switchboard).

## **Peri-operative Management of Patients with Known Sickle Cell Disease**

*(NB This includes HbSS, HbSC, Hb SBthal, HbSE and HbSD)*

### **A. Pre-operative management (elective surgery)**

All patients with Sickle Cell Disease who need general anaesthesia and surgery should be discussed with the sickle cell team, who will organise to review the patients and will produce a pre-operative plan.

Surgical team:

- Should contact sickle cell team via letter or email when they list patient for surgery to allow time for haematology review.

Pre-assessment team:

- Any patient known to have sickle cell disease (or diagnosed de novo as part of pre-operative screening) should be discussed with the sickle cell team (via phone or email) to confirm the pre-operative plan
- Organise appropriate pre-operative blood tests (Table 1). Other routine tests should also be taken as per usual pre-assessment protocol

Table 1

Investigation	Reason
Full blood count	Compare Hb with previous results.
Haemoglobin electrophoresis	If this has not previously been done.
Renal function	To assess for sickle nephropathy
Group and antibody screen	To confirm there are no red cell antibodies
Extended RBC phenotype	If this has not previously been done

Sickle cell team:

- Will review all pre-operative patients in the sickle outpatient clinics (Tues pm and Friday am)
- Assess any previous analgesia requirements in order to avoid respiratory depression and inadequate pain control
- Assess end-organ damage, for example renal damage, pulmonary hypertension and cardiomegaly.
- Order echo if not performed within previous 2 years. Review baseline saturations and sleep study if available.
- Refer for specialist advice if concerns re cardio-pulmonary complications or other end organ damage.
- Refer for anaesthetic review if concerns about medical co-morbidities or pain control.
- Discuss need for pre-operative blood transfusion and organise
- Enter transfusion plan on 'Pre-operative Assessment MDM Outcome' on EPR and inform pre-assessment and surgical team re transfusion plan.
- Will review post-operative patients daily whilst in-patients

## **Pre-operative transfusion**

Patients with sickle cell disease have increased risks of peri-operative complications which can be decreased by appropriate pre-operative blood transfusion.

### **HbSS and HbS<sup>0</sup>thalassaemia :**

**All patients with HbSS and HbS<sup>0</sup>thalassaemia should be offered pre-operative transfusion 2-10 days prior to surgery.**

Minor or moderate risk operations (D&C, tonsillectomy, splenectomy, arthroscopy, laparoscopic surgery):

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 (neurosurgery, cardiac surgery or complex orthopaedic surgery, including total hip replacement): Exchange blood transfusion 2-10 days prior to surgery aiming for pre-op Hb of 100g/l and HbS% of <60%.

### **HbSC and other genotypes.**

There is less clear evidence for role of transfusion in this group of patients. Therefore the haematology will review each case on a case by case basis and decide on treatment depending on operation and disease severity. In general:

Minor or moderate risk operations (D&C, tonsillectomy, splenectomy, arthroscopy, laparoscopic surgery):

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

Hb >90g/l – no transfusion, unless very severe disease phenotype.

*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 (neurosurgery, cardiac surgery or complex orthopaedic surgery, including total hip replacement): Exchange blood transfusion 2-10 days prior to surgery aiming for pre-op Hb of 100g/l and HbS+C% of <60%.

## **Other transfusion issues**

1. The blood transfusion lab should always be informed via the request form that the blood is for a patient with Sickle Cell Disease. All patients with Sickle Cell Disease should be given full Rh (C, D and E) and Kell matched blood. An extended red cell phenotype should be taken before giving blood. Blood should be HbS negative.

2. Many patients with Sickle Cell Disease have received multiple transfusions and many have multiple red cell antibodies. It is important therefore to give Blood Transfusion 24 hours notice of transfusion requirements since it may be necessary to obtain compatible blood from the Regional Transfusion Centre.

3. In some cases patients are 'untransfusable' because of multiple red cell antibodies or because of hyperhaemolysis (an immunological complication of red cell transfusion). In these cases transfusion requirements should always be discussed with the sickle cell

team. These patients may require prolonged treatment with erythropoietin pre-operatively to increase their Hb level.

See 'Management of Patients Refusing Blood Transfusion Including Jehovah's Witness Patients' and 'Guidelines for the management of hyperhaemolysis in patients with Sickle Cell Disease, including the use of intravenous immunoglobulins (IVIg)'

## **B. Pre-operative management (emergency surgery)**

All patients with Sickle Cell Disease who need general anaesthesia and surgery should be discussed with the sickle cell team/on call haematology team out of hours, who will organise to review the patient and will produce a pre-operative plan.

Surgical team:

- Should contact sickle cell team/on call haematology team out of hours to discuss patient with them
- Ask anaesthetic team to review
- Organise appropriate pre-operative blood tests (Table 1). Other routine tests should also be taken as per usual pre-assessment protocol

Table 1

Investigation	Reason
Full blood count	Compare Hb with previous results.
Haemoglobin electrophoresis	If this has not previously been done.
Renal function	To assess for sickle nephropathy
Group and antibody screen	To confirm there are no red cell antibodies
Extended RBC phenotype	If this has not previously been done

Sickle cell team/on call haematology team:

- Confirm sickle status/genotype
- Review Hb/organ function and develop pre-operative plan
- Ensure no history of transfusion complications/red cell antibodies.
- Refer for specialist advice if concerns re cardio-pulmonary complications or other end organ damage.
- Discuss need for pre-operative blood transfusion with on call haematology and discuss with surgical team.
- Review patient on ward pre-operatively if there is time prior to surgery, or post-operatively to review analgesia/oxygenation.

### **Pre-operative transfusion**

Patients with sickle cell disease have increased risks of peri-operative complications which can be decreased by appropriate pre-operative blood transfusion. In the emergency situation decisions will be based on the sickle genotype, Hb level, urgency of surgery, type of surgery and pre-morbid condition of patient.

General guidance

- Most patients with HbSS and HbSB<sup>0</sup>thalassaemia will have Hb <90g/l and can be managed with a top up transfusion. Exceptions are: high risk surgery

or significant chronic disease when exchange transfusion may be indicated, or patients with mild phenotype and Hb >90g/l when surgery should take place without transfusion

- Most patients with HbSC will have mild disease and Hb >90g/l and surgery should take place without transfusion. Exceptions are: high risk surgery, Hb <90g/l or patients with significant chronic disease when transfusion should be considered
- If surgery is life-threatening and urgent, it should be performed and blood transfusion (top up or exchange) can be given intra-operatively or post-operatively.

### **C. Intra-operative management**

Start iv fluids once patient is nil by mouth. Patients with sickle cell disease usually have urinary concentrating defects so can become rapidly dehydrated. Continue until patient is drinking freely.

- No specific anaesthetic technique is recommended.
- Full monitoring
- Pre-oxygenation
- Positioning to avoid venous stasis
- Measures to avoid heat loss

### **D. Post-operative management:**

- Continue pulse-oximetry (on air). Call Haematology team if <94%.
- Continue iv fluids until drinking freely
- Care in recovery or HDU or ITU for at least 24 hours if possible for major operations without exchange blood transfusion.
- O<sub>2</sub> therapy for 48 hours to maintain oxygen saturations >94% (major cases)
- Consider ventilatory support with incentive spirometry or continuous positive airways pressure
- Effective analgesia -. Many patients with SCD are opiate naïve and will respond to normal post-operative pain management. A small number of SCD are opiate tolerant and may require higher than normal levels of analgesia. Each patient should be evaluated individually and their normal sickle cell inpatient and out patient pain regimes should give an indication of post-operative pain requirements. Opiate tolerant patients may need discussion with the acute pain service and the sickle cell team before surgery so that an appropriate pain management regime can be prescribed and implemented following surgery.
- Normothermia
- Consider thromboprophylaxis with dalteparin prophylactic doses.
- Continue penicillin V, folic acid and other regular medications (including hydroxycarbamide) unless there is a contra-indication.
- DO NOT use ice packs to reduce swelling.

## **E. Regional anaesthesia in sickle cell disease:**

Sickle complications are less likely with spinal or epidural anaesthesia than with general anaesthesia and unless patients have very severe phenotypes or are very anaemic (<65-70-g/l ), patients are unlikely to need pre-operative transfusion.

Homozygous sickle cell anaemia is usually regarded as a contraindication to the use of a tourniquet, despite anecdotal reports of uneventful use in such patients. Risks of complications following tourniquet use may be diminished if pre-operative transfusion is used. There is little evidence about the use of tourniquet in sickle cell trait, but it is probably appropriate to keep tourniquet time to a minimum in these patients. In cases of prolonged tourniquet time, or where there are special concerns it maybe worthwhile instituting intra-operative measures as for sickle cell disease patients.

### **Patient screening: Sources of information and Support**

If the patient has been found to have found to have sickle trait or newly diagnosed Haemoglobinopathy and they have concerns or questions about this they should be encouraged to discuss this with their GP. General patient information about sickle cell trait or disease (in those cases who are newly diagnosed) can also be obtained from the sickle cell society [www.sicklecellsociety.org](http://www.sicklecellsociety.org). The sickle cell society can be also be contacted on 02089617795 and they can also advise patients with concerns or questions about their local sickle cell services and counsellors

### **Monitoring of this protocol:**

Clinicians who encounter problems implementing this protocol in individual cases should discuss them with duty haematology staff. Unresolved issues should be reported to Dr Jo Howard, Consultant Haematologist X82741, or Dr Rachel Kesse-Adu, Consultant Haematologist, Laboratory haematology X82736 (haematology staff) or Dr Andrew Morley, Consultant Anaesthetist X80652 (other clinicians). Recurring difficulties will be discussed between Drs Morley and Howard and presented at departmental audit meetings if appropriate.

### **References:**

Fisher B, Roberts CS. Tourniquet use and sickle cell haemoglobinopathy: how should we proceed? *South Med J.* 2010 103 (11): 1156-60

Howard J, Malfroy M, Llewelyn C, Choo L, Hodge R , Johnson T, Purohit S, Rees DC, Tillyer L, Walker I, Fijnvandraat K , Kirby-Allen M, Spackman E, Davies SC, Williamson LM. Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS): a randomised controlled multi-centre clinical trial : *Lancet* 2013, 381 (9870) 930-8

Vichinsky EP, Neumayr LD, Haberkern C, et al. A comparison of conservative and aggressive transfusion regimens in perioperative management of sickle cell disease. The Perioperative Transfusion in Sickle Cell Disease Group. *N Engl J Med.* 1995; 333: 206 – 213.

## **APPENDIX 1**

### **Adult sickle cell team**

**Department of Haematology, 4<sup>th</sup> Floor, Southwark Wing, Guy's Hospital  
Great Maze Pond, SE1 7EH: 02071887188**

### **Medical team**

**Consultants: Dr J Howard / Dr R Kesse-Adu – ext 81432/82741**

**PA to consultants, Ms Anne Oddotte ext 82741**

**Specialist Registrar- Bleep 0248**

**SHO - Bleep 2283**

### **Nursing team**

#### **Advanced Nurse Practitioner**

**Mr Neill Westerdale- Bleep1843/ Ext 82710/ Mob 07770683947**

#### **Clinical Nurse Specialist (Teenagers and Young Adults)**

**Mr Luhanga Musumadi. Bleep 2256/Ext 82710/ Mob 07770678851**

#### **Clinical Nurse Specialist**

**Ms Judith St Hilaire - Ext 82710/ Bleep 2868 / Mob 07920711266**

### **Out of hours**

**Haematology SpR on call via switch (02071887188)**



## APPENDIX 2

### Contributors to protocol

Dr Jo Howard	Consultant Haematologist	Clinical Haematology, Oncology and Haematology Directorate
Dr Rachel Kesse-Adu	Consultant Haematologist	Clinical Haematology, Oncology and Haematology Directorate
Mr Neill Westerdale	Advanced nurse practitioner: sickle cell disease	Clinical Haematology, Oncology and Haematology Directorate
Mr Luhanga Musumadi	Clinical Nurse Specialist: Transition / haemoglobinopathies	Clinical Haematology, Oncology and Haematology Directorate
Mrs Judith St Hilaire	Clinical Nurse Specialist. Haemoglobinopathies	Clinical Haematology, Oncology and Haematology Directorate
Dr Andrew Morley	Consultant anaesthetist	Perioperative medicine directorate
Dr Charles Thoburn	Consultant anaesthetist	Perioperative medicine directorate
Dr Nicky Thomas	Consultant health/clinical psychologist	Clinical Haematology, Oncology and Haematology Directorate
Dr Yvonne Daniels	Chief biomedical scientist	Laboratory Haematology, Diagnostic/Therapeutic Directorate