



**SPAHA**

SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

## Scottish Paediatric & Adult Haemoglobinopathy Network

### Perioperative Management of Adult Patients With Sickle Cell Disease

#### **NOTE**

This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

## Principles of management

The Haematology Team should be informed as far in advance of elective surgery as possible. Preparation for surgery will usually require **AT LEAST two week's notice**.

**For emergency admissions inform the Haematology Team and Blood Transfusion Laboratory immediately if surgery is deemed likely.**

Patients should be managed jointly between haematology, surgery and anaesthetics. The Anaesthetic Team should be made fully aware of the patient's needs.

An individual management plan should be in place prior to surgery which takes into account the patients' particular risk factors and the type of procedure planned. This should be circulated to all relevant individuals prior to an elective surgical procedure.

If a period in HDU/ICU is anticipated the necessary arrangements should be made in advance.

Sickle patients should be placed near the beginning of the theatre list to minimize time fasting and reduce likelihood of cancellation.

Ensure the patient is well informed and involved in their management plan when possible.

Fluid balance must be managed with great care in sickle patients as they dehydrate easily due to impaired renal concentrating ability.

These patients may have a history of chronic pain and be relatively insensitive to opiate analgesia. This should be taken into account when prescribing in the postoperative period.

Avoid factors which may precipitate the development of a sickle crisis;

**Hypoxia**

**Dehydration**

**Acidosis**

**Cold**

**Pain**

Day case surgery may not be suitable for this patient group due to the above considerations.

## Preoperative Care

Ensure Haematology Team is made aware of admission as soon as possible.

Blood transfusion is not always indicated prior to surgery (see Page 4). Where indicated blood transfusion will be arranged by the Haematology Team and arranged during the week before surgery for elective procedures. **Do not transfuse without prior discussion with haematology.**

**In addition to FBC, HbS level and routine bloods, a sample for Group and Save, antibody screening and red cell phenotyping (if not performed previously) should be sent at the pre-op assessment visit and BTS informed of the date and nature of the planned procedure.**

Admit the day before surgery if possible.

### Hydration

Maintenance IV fluids should be commenced the evening before surgery unless the patient is drinking freely.

IV fluids must be started if nil by mouth for >2 hours.

Maintenance fluids may need to run at a higher rate than usual to avoid dehydration. Monitor fluid balance closely.

### Thromboprophylaxis

This should be considered for all procedures, particularly for major surgery or if patient will be immobile for >24 hours post procedure.

Follow local guidelines on VTE prophylaxis.

### Oxygenation

Avoidance of hypoxia is vital to prevent sickling and tissue ischemia.

Document baseline O<sub>2</sub> saturation (may have pre-existing cardiorespiratory disease)

Consider use of incentive spirometry.

Oxygen saturation should be monitored continuously once premedication given.

Consider giving supplemental oxygen from the time of premedication.

Hyper-oxygenate at induction of anaesthesia.

### Temperature Regulation

Hypothermia can trigger peripheral stasis and sickling.

Attention should be paid to keeping the patient normothermic during surgery. This may require the use of warmed intravenous fluids, warm air blankets and adjustment to the ambient temperature in theatre. Ensure the anaesthetic team is made aware of these requirements in advance.

### Infection Management

These patients are functionally hyposplenic and are therefore at increased risk of perioperative infection. If additional antibiotic prophylaxis is needed this should be detailed in the patient's management plan.

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

## Intraoperative Care

### Hydration

Volume status must be monitored closely throughout the procedure and hypovolaemia avoided at all costs through the prompt use of volume replacement. Sickle patients are more prone to dehydration due to impaired renal concentrating ability.

### Positioning

As prolonged venous stasis can lead to sickling, the patient should be repositioned periodically where possible during long procedures.

### Tourniquet

These are generally contraindicated in patients with sickle cell disease and relatively contraindicated in sickle cell trait.

### Oxygenation

Continuous pulse oximetry throughout the procedure.

### Temperature Regulation

Ensure the patient remains normothermic. Warmed IV fluids are particularly important during long procedures.

### Cell Salvage

Intraoperative cell salvage is contraindicated in patients with sickle cell disease.

## Postoperative Care

### Hydration

Continue IV fluids until the patient is able to tolerate a sufficient oral intake. Avoid fluid overload post-op as this may increase the risk of complications in high risk patients.

### Analgesia

Prescribe post-op analgesia as per local guidelines. 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-op period (see separate guideline).

### Oxygenation

Pulse oximetry should continue for at least 24 hours or until stable. Maintain saturations >95% or above baseline whichever is higher. Continue incentive spirometry, encourage patient to take regular deep breaths (every 10 minutes) if not available.

### Temperature regulation

Ensure the patient is normothermic until able to regulate body temperature independently.

### Infection Management

If patient develops a fever (>38.0°C) ensure blood cultures are taken and start intravenous antibiotics as per local antimicrobial policy (NB hyposplenic).  
Check cannula and line sites daily for signs of infection.  
Monitor for symptoms such as productive cough, shivering or myalgia.

### Thromboprophylaxis

This should be considered for all procedures, particularly for major surgery or if patient will be immobile for >24 hours post procedure.  
Follow local guidelines on VTE prophylaxis.  
Encourage early mobilisation with physiotherapy input if appropriate.

**Should the patient develop new symptoms suggestive of a sickle related complication, such as a painful crisis, chest crisis or CVA the Haematology Team MUST be informed immediately.**

## Blood Transfusion Guidance

### Transfusion Prior to Elective Surgery

Patients undergoing major surgery and those with a history of previous sickle related complications are at highest risk in the perioperative period. Conversely over aggressive transfusional support may unnecessarily expose low risk patients to transfusion related hazards. Transfusion should therefore be considered on a case by case basis dependant on patient factors and the nature of surgery.

In patients with a high baseline Hb of >90g/l top-up transfusion should generally not be used as this may precipitate hyperviscosity. **Care should be taken not to exceed baseline Hb values for sickle cell patients with high steady state Hb(>100 g/l).**

***Particular caution is needed in patients with HbSC disease as they are more likely to develop hyperviscosity related complications with top-up transfusion.***

Blood should be sickle negative and matched for Rh and Kell antigens as a minimum. **Units should also be negative for any red cell alloantibodies present.**

The TAPS<sup>1</sup> trial has shown benefit for pre-operative transfusion in HbSS patients undergoing low and medium risk surgery. See appendix for comprehensive list of surgical procedures classified by risk<sup>1</sup>. Numbers of patients included with low-risk procedures was small but transfusion should still be considered. Likewise few patients were included in the pre-operative exchange transfusion arm but this should be considered for those with a baseline Hb>90g/l. The trial excluded patients with HbSC disease and S/β<sup>+</sup>, but in most cases it would be reasonable to follow the principles below:

### 1. Low -risk surgery

Top-up transfusion should be considered (irrespective of HbS level) in most cases. Aim for a pre-op Hb of approximately 100g/l. Avoid top-up if baseline Hb>90g/l. Post transfusion Hb should not exceed 100g/l or 10-20g/l above baseline. Transfusion not routinely indicated for patients with uncomplicated HbSC disease.

## 2. Medium-risk surgery

**Transfusion indicated for both HbSS and HbSC disease.**

**Top-up transfusion aiming for Hb of 100g/l, or partial exchange (see Exchange Transfusion guideline) if baseline Hb >90g/l (target HbS <60%). Post transfusion Hb should not exceed 100g/l or 10-20g/l above baseline.**

## 3. High-risk surgery

High-risk procedures e.g. thoracic, major upper abdominal surgery, neurosurgery or previous serious sickle related complications e.g. severe chest crisis, CVA.

**Exchange transfusion aiming for a pre-op HbS % of <30 (see Exchange Transfusion guideline) for all patients unless a specific contra-indication exists. Hb should be kept below 110g/l.**

The above principles are also applicable to patients undergoing **emergency surgery if time permits**. Emergency surgery should not be delayed solely to permit transfusion and intra-operative or post-operative transfusion should be considered as an alternative. As a minimum these patients should have suitable cross-matched blood available should an emergency exchange procedure be needed (see Exchange Transfusion guideline).

### Transfusion reactions/haemolysis

- Sickle patients are at increased risk of delayed haemolytic transfusion reactions which can be very severe, so called 'hyperhaemolysis.'
- These can typically occur around one week post-transfusion but may occur sooner in those with a history of previous transfusion reactions.
- If the patient develops an unexpected drop in haemoglobin and becomes jaundiced liaise with haematologist to see if any further investigations required to exclude haemolysis. Postpone further transfusion until discussed.

## References

1. Howard J, Malfroy M, Llewelyn C et al. The Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS) study: a randomised, controlled, multicentre clinical trial. Lancet 2013; 381: 930–38.
2. Davis B, Allard S, Qureshi A et al. Guidelines on Red Cell Transfusion in Sickle Cell Disease Part I: Principles and laboratory tests. BJH 2017; **176**: 179-191
3. Davis B, Allard S, Qureshi A et al. Guidelines on Red Cell Transfusion in Sickle Cell Disease Part II: Indications for Transfusion. BJH 2017; **176**: 192-209

## Appendix 1

### TAPS SURGICAL RISK SCORES

Surgery Type	High Risk	Medium Risk	Low Risk
<b>General</b>			
Biliary Surgery		Medium	
Cholecystectomy and exploration of common duct		Medium	
Colostomy		Medium	
Gastrostomy		Medium	
Hepatectomy	High		
Ileostomy		Medium	
Laparotomy		Medium	
Liver Biopsy		Medium	
Mastectomy		Medium	
Oesophagectomy	High		
Oesophagogastrrectomy	High		
Partial Gastrectomy		Medium	
Splenectomy		Medium	
Total Gastrectomy		Medium	
Vagotomy +/- drainage		Medium	
<b>Endocrine</b>			
Adrenalectomy		Medium	
Parathyroidectomy		Medium	
Partial Pancreatectomy		Medium	
Thyroidectomy		Medium	
Whipples procedure	High		
<b>Transplantation</b>			
Bone Marrow Harvest			Low
Kidney transplant		Medium	
Colo-Rectal Surgery			
Intra-abdominal - colectomy		Medium	
Rectopexy		Medium	
Rectum - pouch; resection/excision		Medium	
<b>Vascular Surgery</b>			
Amputation of limb		Medium	
Aorto-femoral bypass	High		
Aorto-iliac endarterectomy	High		
Axillo-pop bypass		Medium	
Bifemoral bypass		Medium	
Endarterectomy (HIGH if carotid/vertebrobasilar)		Medium	
Fem-pop bypass		Medium	

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<b>Surgery Type</b>	<b>High Risk</b>	<b>Medium Risk</b>	<b>Low Risk</b>
Infra-renal aneurysm		Medium	
Sympathectomy		Medium	
Thoracic Aortic Aneurysm	High		
<b>Cardiothoracic Surgery</b>			
Angioplasty	High		
Aortic Valve Replacement (AVR)	High		
Coronary Angioplasty	High		
Coronary Artery Bypass Graft (CABG)	High		
Excision of mediastinal mass	High		
Hiatus Hernia (HIGH in infant)		Medium	
Lobectomy	High		
Mitral Valve Replacement (MVR)	High		
Oesophageal Resection	High		
Pneumonectomy	High		
Re-do AVR	High		
Re-do CABG	High		
Re-do MVR	High		
Sternal deformity	High		
Thorabdominal excision retroperitoneal malig tumour	High		
Thoractomy	High		
Thymectomy	High		
<b>Paediatric Cardiac</b>			
Artrial Septal defect ( ASD)	High		
Blalock Shunt ( simple < 6 years old)	High		
Coarctation < 2 years old	High		
Coarctation 2 - 7 years old	High		
Coarctation 8-15 years old	High		
Other Bypass cases ( e.g. transposition)	High		
Paed ventricular aneurysm	High		
Patent ductus arteriosus (PDA)	High		
Pulmonary artery banding	High		
re-do Blalock for > 6 years old	High		
Re-dos	High		
Unifocalisation	High		
Ventricular septal defect (VSD)	High		
<b>Neurosurgery</b>			
A-V malformations	High		
Craniectomy	High		
Cranioplasty	High		
Craniotomy	High		
Disc Surgery		Medium	
Laminectomy		Medium	
Meningioma	High		
Perpherial Nerve Surgery		Medium	
Posterior fossa exploration	High		



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<b>Surgery Type</b>	<b>High Risk</b>	<b>Medium Risk</b>	<b>Low Risk</b>
Shunt procedures	High		
Spinal decompression	High		
Vascular ( e.g. aneurysm)	High		
<b>Orthopaedics</b>			
Acetabular shelf reconstruction		Medium	
Arthroscopy (shoulder/knee)		Medium	
Bone graft from iliac crest		Medium	
Core decompression of hips		Medium	
Foot or ankle surgery			Low
Incision drainage and curettage of tibia & humerus			Low
Internal fixation of tibia or fibula		Medium	
Intramedullary femoral nail		Medium	
Osteotomy (long bone)		Medium	
Pelvic osteotomy		Medium	
Postop tibia reaming for osteomyelitis			Low
Removal of cervical rib		Medium	
Removal of metal work ( including femoral nail)		Medium	
Repair of Fractured neck of Femur		Medium	
Revision of finger fracture			Low
Revision of Total Hip replacement		Medium	
Revision of total knee replacement		Medium	
Scarf osteotomy 1st metatarsal			Low
Scoliosis Surgery	High		
Soft tissue tumour resection		Medium	
Spinal Fusion (simple)		Medium	
Spinal Fusion complex		Medium	
Total Elbow replacement		Medium	
Total Hip replacement		Medium	
Total Knee replacement		Medium	
<b>Urology</b>			
Augmentation cystoplasty		Medium	
Bladder neck surgery		Medium	
Bladder tumour removal		Medium	
Colpal suspension		Medium	
Cystectomy	High		
Cystoscopy urethral dilation		Medium	
Cystourethrectomy		Medium	
Impospadias repair		Medium	
Nephrectomy		Medium	
Nephropylolithotomy		Medium	
Percutaneous nephrolithotomy (PCNL)		Medium	
Prostatectomy -TURP (HIGH if radical)		Medium	
Pyelolithotomy		Medium	
Renal biopsy			Low
Urethroplasty		Medium	
Urethrolithotomy		Medium	

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Surgery Type	High Risk	Medium Risk	Low Risk
<b>Obstetrics</b>			
ERPC			Low
Hydatidiform mole		Medium	
Hysterectomy ( abdominal or vaginal)		Medium	
LSCS (Caesarian)		Medium	
LSCS for placenta praevia		Medium	
Myomectomy		Medium	
Pelvic extenteration		Medium	
Radical Oophorectomy		Medium	
Radical Vulvectomy		Medium	
Sterilisation		Medium	
Termination of Pregnancy			Low
<b>Maxillofacial</b>			
Bicoronal flap	High		
Bimaxillary osteotomy		Medium	
Craniofacial Surgery	High		
Microvascular		Medium	
Neck Dissection		Medium	
Parotidectomy		Medium	
<b>ENT</b>			
Adenoidectomy & myringotomy			Low
Adenoidectomy & SMD inferior turbinates			Low
Adenoidectomy & submucous diathremy of interior turbinates			Low
Adenoidectomy			Low
Adenoido-tonsillectomy		Medium	
ENT: insertion of stents			Low
Tonsillectomy		Medium	
<b>Eye Surgery</b>			
Trauma/Reconstruction/Orbital surgery		Medium	
Cataract/Virectomy/Retinal surgery			Low
<b>Miscellaneous</b>			
Dental surgery			Low
Drainage of abscess			Low
Excision burns scars			Low
Excision of lipoma			Low
Excision of lymph nodes			Low
Hernia repair (inguinal/umbilical)			Low
Lymph node biopsy			Low
PICC line insertion			Low
Portocath removal/insertion			Low