STSTN Peri-operative management of paediatric patients with sickle cell disease

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Summary

Patients with sickle cell disease are at high risk of experiencing complications, such as painful crisis and acute chest syndrome, when undergoing surgery. This guideline will allow identification and appropriate management of at-risk paediatric patients, in order to reduce peri-operative complications.

1. INTRODUCTION

Sickle disease is a genetic disorder due to a single mutation resulting in the substitution of amino acid valine for glutamic acid in position six of the β-chain of the haemoglobin molecule. This change is responsible for altered haemoglobin with increased viscosity and the development of the 'sickle' shape in deoxygenated states.

The inheritance of a homozygous (SS) or double heterozygous (SC, Sβ-thalassaemia or SD, SE, SOArab) sickle cell disease predisposes the patient to risk of sickling complications, particularly during hypoxia or acidosis.

Sickle cell complications such as painful vaso-occlusive episodes, acute chest crisis and infection are thought to be precipitated by general anaesthesia. It is therefore vital to provide the optimal management of children prior to surgery. Recent studies have demonstrated the benefit of pre-operative transfusion, in order to reduce the rate of peri-operative complications. It is also imperative to avoid cooling, maintain adequate oxygenation, hydration and pain control before, during and after surgery. This guideline aims to allow identification of at-risk patient groups, and guide peri-operative management of paediatric patients with sickle cell disease.

2. PERI-OPERATIVE MANAGEMENT OF PAEDIATRIC PATIENTS WITH KNOWN SICKLE CELL DISEASE

Patients included in this category:

- HbSS/Sβ0thal
- HbSC
- HbSβ+thal
- HbSE
- HbSD
- HbSO Arab

2.1 ELECTIVE SURGERY (Flow chart summary: Appendix 1)

2.1.1. Pre-operative assessment for elective surgery

Table 1

INVESTIGATION	REASON
Full blood count + Reticulocytes	Compare haemoglobin with previous results
Haemoglobin electrophoresis	*If this has not been previously done*
Renal and Liver Profile	To assess for kidney and liver function
Group and antibody screen	To confirm there are no red cell antibodies
Extended RBC phenotype	*If this has not previously been done*

The Surgical team:

- Should contact the sickle team via letter or email when they list patient for surgery to allow time for haematology review.
- For all patients, both the Local and Specialist Haematology/Sickle team must be made aware of the plan for elective surgery (in Specialist centres this will be the same team).

Kings College Hospital: kch-tr.paedhaematologycns@nhs.net
Guys & St Thomas's Hospital: HaemoglobinopathyCNS@gstt.nhs.uk
Evelina London Children's Hospital (GSTT): gst-tr.paediatricsicklethal@nhs.net
Queen Elizabeth Hospital: henrietta.adamah@nhs.net
University Hospital Lewisham: henrietta.adamah@nhs.net
Croydon University Hospital: kch-tr.paedhaematologycns@nhs.net

The Pre-assessment team:

- Should discuss with the sickle cell team (via phone or email) to confirm the pre-operative plan and record this in the notes.
- Should organise appropriate pre-operative blood tests (Table 1). Other routine tests should also be taken as per usual pre-assessment protocol.
- Arrange appropriately cross-matched blood prior to admission for surgery, including those cases in which there is a positive red cell alloantibody.
- Should ensure that all blood for transfusion be fully matched for ABO, Rh phenotype and Kell antigen and any other red cell antibodies previously identified and must be sickle negative.
- Should arrange a post-op HDU/PICU bed for high risk patients undergoing minor/ intermediate surgery (Table 2) prior to admission, which will be in a Specialist Centre.

The Paediatric Sickle Cell team will:

- Be made aware in advance of any plans for surgical intervention for all patients with SCD.
- Discuss all pre-operative patients at the multi-disciplinary team (MDT) meeting.
- Assess any previous analgesia requirements in order to avoid respiratory depression and inadequate pain control – including discussion with anaesthetics.
- Ensure there is documented evidence of transcranial Doppler (TCD) within the last 12 months for children between 2-16 years with HbSS/Sβ0 thalassaemia.
- Wherever feasible, assess end-organ damage, for example renal damage, pulmonary hypertension and cardiomegaly, and request specialist advice if necessary, regarding cardio-pulmonary complications or other end organ damage.
- Discuss the need for pre-operative blood transfusion (see below) and organise for this to take place.
- Enter a transfusion plan on Pre-Operative Assessment MDM Outcome on the electronic patient record (Appendix 3) and inform the pre-assessment and surgical team re transfusion plan.
- **If the patient is not routinely managed in the operating team's Specialist Haemoglobinopathy Centre, all the above information must be sent by the patient's Local Haemoglobinopathy or usual Specialist Centre to the sickle team in where surgery is to take place, via letter or email at least 4 weeks before surgery.**

2.1.2. General considerations in pre-operative management for elective surgery

Children with SCD are not considered suitable for day case surgery:

- They should be listed as early as possible on the operating list.
- Maintenance IV fluids should commence from time of becoming nil by mouth and continue until the child is drinking freely.
- Oxygen saturation should be maintained above 96% and encourage 2-hourly incentive spirometry/bubble therapy when they are fully awake.
- Pain: ensure adequate pain control and consider using iv/po morphine post anaesthesia, with appropriate monitoring of pain score and sedation.
- Avoid using tourniquets and avoid use of cell salvage for these patients.

2.1.3. Pre-operative transfusion for elective surgery

The risk of general anaesthesia is shown to be mitigated by appropriate patient selection and the use of blood products prior to surgery.
See Table 2: Assessment of Risk of Surgical Procedures: Low, Intermediate and High Risk depending on Procedure and patient factors.

Table 2
Assessment of Risk of Surgical Procedures: low, intermediate and high risk depending on the procedure and patient factors.

Low Risk

Patient: no previous severe complications

AND

Low risk surgery e.g. simple dental extraction, grommets, hernia repair, orchidopexy.

- Simple top-up transfusion should be considered on an individual basis if Hb<90g/l, aiming for pre-op Hb 100g/l for HbSS and HbSβ^o patients.

- Patients with HbSC disease usually have higher steady state Hb (>90g/l) and top up is likely to cause hyper-viscosity and complications.

Intermediate Risk

Patient: previous history of Acute Chest Syndrome (or other co-morbidity)

AND

Low risk surgery e.g. simple dental extraction, grommets, hernia repair, orchidopexy.

OR

Patient: no additional patient risk factors

AND

Intermediate risk surgery e.g. tonsillectomy, adenoidectomy with obstructive sleep apnoea, splenectomy or cholecystectomy, Portacath insertion.

- Simple top-up transfusion, target Hb of 100g/l.
- In patients with a high baseline Hb (>90g/l) or HbSC disease, top-up transfusion should be avoided due to risk of hyperviscosity; consider exchange transfusion.

High Risk

Patient: previous life-threatening sickle-related complications, e.g., severe ACS, CVA or PICU admissions

AND/OR

High risk surgical procedures e.g. cardiothoracic, major abdominal surgery, neurosurgery, posterior chamber ophthalmic surgery, joint replacement, complex orthopaedic surgery, organ transplantation

- Red cell exchange transfusion aiming for HbS <30% or HbS + C% <30% and pre-op Hb 100g/l; either a manual or automated exchange procedure within a week of planned surgery, depending on patient's size, availability of apheresis etc.

- For patients on regular transfusions with HbS <30%, either a simple top up transfusion can be arranged, or the timing of their scheduled transfusion can be organised 2-7 days before planned surgery.
- Organize an HDU/PICU post-op bed in discussion with the surgeon, anaesthetist and HDU/PICU team

2.1.4. Post-operative management for elective surgery

- For high risk patients undergoing minor/intermediate surgery (Table 2) post-operative care should take place in HDU/PICU at KCH/ECH.
- Pulse Oximetry should be recorded regularly and supplementary oxygen administered where necessary to ensure SpO2 > 96%.
- Avoid dehydration and give maintenance IV fluids until able to drink adequately.
- Pain control: consider using IV morphine and where necessary PCA / NCA (patient or nurse control anaesthesia).
  The standard analgesia should be given regularly and not as PRN, in order optimise pain control.
- If the patient develops a fever, they need assessment and antibiotics started promptly. Stop regular Penicillin V but remember to restart when course completed.
- Consider thromboprophylaxis in post pubertal patients, particularly those with a high BMI, prolonged immobility or previous VTE, unless contraindicated.

2.2. EMERGENCY SURGERY

2.2.1. Pre-operative management for emergency surgery

Surgical team:

- Should contact sickle cell team/on call haematology team out of hours to discuss patient with them. If an LHT patient, please contact on call SHT haematology team to discuss patient.
- Should ask the anaesthetic team to review pre-operatively.
- Should organise appropriate pre-operative blood tests (Table 1). Other routine tests should also be taken as per usual pre-assessment protocol.

Paediatric Sickle Cell team/on call haematology team will:

- 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.

2.1.2. General consideration in pre-operative management for emergency surgery

- Maintenance IV fluids should commence from time of nil by mouth and continue until the child is drinking freely.
- Oxygen saturation should be maintained above 96% and encourage 2-hourly incentive spirometry or bubbles for children when they are fully awake.

2.2.3. Pre-operative transfusion for emergency surgery

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 situations decisions will be based on the sickle genotype, Hb level, urgency of surgery, type of surgery and pre-morbid condition of patient.
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.

Please refer to section 2.1.3 re risk stratification

3. MANAGEMENT OF PAEDIATRIC PATIENTS WITH KNOWN SICKLE CELL DISEASE UNDERGOING GENERAL ANAESTHESIA FOR RADIOLOGICAL PROCEDURES SUCH AS MRI/MRCP/ERCP

- Contact LHT or SHT sickle cell team to discuss patient's history and determine procedure.
- Transfusion generally not required unless history of severe ACS, severe cerebrovascular disease or severe anaemia.
- Give maintenance IV fluids while nil by mouth until drinking normally.

4. **PATIENTS DUE TO HAVE SPLENECTOMY**

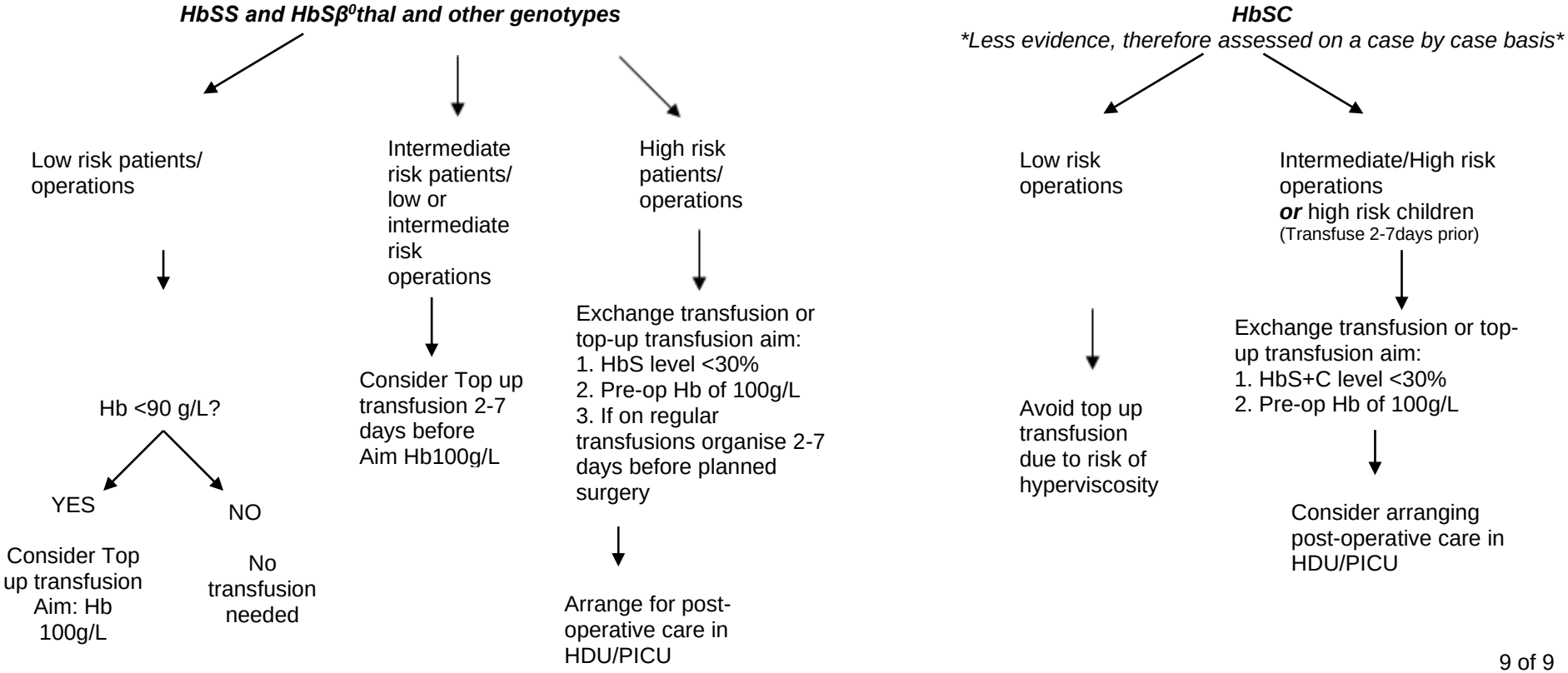
- Please ensure all patients have had required vaccines at least 4 weeks before surgery: Pneumovax, given age 2 years and then every 5 years, Hib booster and Meningitis ACWY. Confirm with the SHT centre where surgery being carried out, either by letter or email.

5. REFERENCES

1. Howard et al, The Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS) study: a randomised, controlled, multicentre clinical trial. Lancet 2013; 381: 930-38
2. Vichinsky et al A comparison of conservative and aggressive transfusion regimens in the perioperative management of sickle cell disease NEJM vol 333 no4 206 –213
3. Fullerton MW, Philippart AI, Sarnaik S, Lusher J. Preoperative exchange transfusion in sickle cell anaemia. J Ped Surg 1981; 16:297-300.
4. Bischoff RJ, Williamson A III, Dalali MJ, et al. Assessment of the use of transfusion therapy perioperatively in patients with sickle cell haemoglobinopathies. Ann Surg 1988;207:434-8

APPENDIX 1: PRE-OPERATIVE TRANSUFION IN SICKLE CELL: ELECTIVE SURGERY

Surgical team to inform sickle team of date of surgery
Preoperative assessment team to perform pre-operative blood tests
(FBC, retics, Hb electrophoresis, U&Es, LFTs, G&S, Extended RBC phenotype)
Sickle team to discuss at MDM and make pre-operative transfusion plan:
Please refer to table 2 in section 2.1.3 for risk assessment



APPENDIX 2:

Proforma: Pre-Operative Checklist to be sent to specialist centre

To be filled in by the sickle cell team – then to be uploaded onto electronic record and copy to be placed in the notes

Patient details:

PAST MEDICAL HISTORY

Sickle genotype:

Comorbidities: ?ACS, stroke, PICU

Medications:

Allergies:

Patients family Jehovah's witness?

Date of last TCD: Normal Abnormal Conditional Inadequate (Circle)

TRANSFUSION HISTORY

Baseline Hb (g/L):

Known red cell antibodies? ☐ Yes ☐ No If yes, which Ab?

On transfusion programme? ☐ Yes ☐ No If yes, indication?

Previous severe transfusion reactions:

PLANNED PROCEDURE/IMAGING under GA

Type of Surgery/Imaging:

Hospital:

Date of surgery:

PRE-OPERATIVE PLAN

To be discussed at the paediatric sickle cell MDM

For discussion with anesthetics pre-op? ☐ Yes ☐ No

For PICU/HDU post-operatively? ☐ Yes ☐ No

Pre-operative transfusion required? ☐ Yes ☐ No

Please refer to peri-operative management of sickle cell disease in paediatric patients protocol 2020

If yes: Target Hb (g/L) or Hb S / HbC%

Date of transfusion

Additional notes:

Checklist sent to SHT

Send to email address kch-tr.paedhaematologycns@nhs.net for KCH,

Paediatricsicklecell@gstt.nhs.uk for ECH

Plan to be organised by: Sickle team member:

Contact details:

Contact details:

KCH Paed sickle CNS: 02032994752
KCH Paed sickle consultants: david.rees2@nhs.net subarna.chakravorty@nhs.net
KCH Paed Sickle Consultant: sue.height@nhs.net
GSTT Paed sickle CNS: 0207 188 9432
GST Paed sickle consultants: Baba.Inusa@gstt.nhs.uk maria.pelidis@gstt.nhs.uk
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UHL Paed sickle CNS: 07741233556
UHL Paeds sickle consultants: s.wilkinson6@nhs.net
CUH Paeds sickle CNS: 0208 2517229
CUH Paeds sickle consultants: nazmachowdhury@nhs.net

Additional contacts can be found on the STSTN website (www.ststn.co.uk)

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