Anaesthesia in Children with Sickle Cell Disease

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Intended Audience

This guideline is intended for staff working at Sheffield Children’s NHS Foundation Trust (SC(NHS)FT) when managing patients with Sickle Cell Disease in whom surgery is planned, or is needed as an emergency.

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1 Introduction

Sickle Cell Disease includes HbSS (sickle cell anaemia), HbSC and HbS/Beta thalassaemia. Patients may have a normal Hb level and may be asymptomatic but are still at risk of sickle related complications.

- All SCD patients are at increased risk of sickle complications during the peri-operative period
- Good communication between surgeon, anaesthetist and haematology team is needed
- A written management plan (see M3/Form1/1096 - Peri-Operative plan for Patients with Sickle Cell Disease) should be agreed and uploaded to the notes prior to surgery. This is a standard suitable for audit. This plan should be triggered by the surgical team informing the haematology team of the patient being listed for theatre.
- Always note sickle cell disease under ‘Additional Information’ on the green booking form. An HDU bed may be needed post-operatively.

If you arrange surgery for a child with sickle cell disease you must contact the haematology team so a management plan can be put in place

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           Haematology Nurse Specialists  Shaun Emmitt Ext 17329, bleep 123
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           Haematology Consultant on call (if out of hours) via switchboard
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2 Pre-Operative Screening

Children born in the England and Wales are likely to have had Newborn Screening for Sickle Cell Disease. The Child’s ‘Red Book’ should be consulted for New Born Screening results.

If there is no documentary evidence of a NBBS outcome in the Red Book, the anaesthetist can contact Angela Ford, Team Leader for the Child Health Service (telephone 3053290) and, through the child’s NHS number, request the information.

Child Health Service is only available from Monday to Friday from 9am to 5pm, and is not available on Bank Holidays or weekends.

If this information cannot be obtained, or the child did not have Newborn Screening (born elsewhere or screening declined) then proceed as below:

All patients of non north European origin should be screened for sickle haemoglobin when surgery is planned. If the sickle solubility test is positive (indicates the presence of sickle haemoglobin, but not whether trait or disease) the haemoglobin phenotype should be determined by haemoglobin electrophoresis or HPLC.

These tests may give false negative results, if

- <4 months has elapsed after a blood transfusion or
- in babies <6/12 old.

However sickle-related problems are unlikely if the sickle solubility test is negative.

Ideally babies of non north European origin <6/12 old due to undergo major surgery should have full Haemoglobin analysis. In reality this may be difficult because of previous transfusions. If born in the UK they are likely to have undergone neonatal screening and the parents will probably know their sickle status. However if there is any doubt it should be checked.

For elective surgery the sickle cell status should have been checked at the pre op clinic. Check that the result is available.

For acute emergencies during normal working hours (Mon-Fri, 9 - 5pm): take an EDTA sample for FBC and film, reticulocytes, sickle solubility test and HPLC.

Out of Hours: take the sickle screening sample (EDTA) for FBC, film and reticulocytes and sickle solubility test to the Haematology laboratory, and contact the Haematology BMS. If the sickle solubility test is negative and the patient is >6/12 old, proceed as normal. HPLC can then be completed the next working day.

If the sickle solubility test is positive, or the patient is <6/12 old and a definitive result is essential, further tests will be necessary; either the surgery must be postponed until the result is known or the patient managed as if they have SCD pending the result.
3 Peri-Operative Management

Sickle Cell Trait (HbAS) patients.

A sickle crisis may be caused by severe local or general hypoxia and in theory the use of an orthopaedic tourniquet could precipitate this; however, experience to date does not indicate an increased risk and if there is a good clinical indication it is considered reasonable to use a tourniquet.

Sickle Cell Disease Patients (SCD)

Schedule SCD patients early on the list so they are not cancelled.

Assess each patient on an individual basis, taking account of pre-existing end organ damage such as CVA, renal or pulmonary disease which may influence management.

Post-operative HDU care is recommended following major surgery.

Optimal management involves attention to the following:

- **Maintain hydration**
  Encourage clear fluids orally (‘Sip until sent for’) prior to induction of anaesthesia, or start intravenous fluids at 1.5 x normal maintenance from ‘Nil by Mouth’ and continue post-operatively until the patient can maintain a good oral intake. SCD causes an inability to concentrate urine properly and can lead to dehydration.

- **Prevent peri-operative cooling**
  This can lead to peripheral stasis in the early post-operative period resulting in tissue hypoxia which may lead to crisis - it is important to prevent this.

- **Prevent hypoxia**
  Pre-op: consider oxygen from the time of pre-medication. Continuous oxygen saturation monitoring is necessary if a respiratory depressant is used. The patient must be pre-oxygenated prior to induction.
  
  Post-op: consider oxygen continuously for 24hrs or until the patient starts mobilising. Consider oxygen on 2nd and 3rd post-op. nights after thoracic or abdominal surgery. Oxygen saturations should be monitored continuously. Arrange chest physiotherapy and incentive spirometry after major surgery.

- **Prevent acidosis**
  If appropriate assess blood gases. The cause of any acidosis should be sought and treated. Discussion with PICU staff may be needed.

- **Manage pain**
  Take all available steps to avoid pain as it may precipitate a sickle crisis.
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- **Transfusion**
  All patients with SCD should have a Blood Group and extended red cell phenotype & antibody screen (2.5mls, EDTA) when put on the surgical waiting list and the blood bank informed of their sickle cell status (hopefully we will already know the patient and this information may already be available – check with blood bank staff). Some patients have red cell allo-antibodies following multiple transfusions and may require specially selected blood; allow at least 48 hours notice if blood is likely to be needed for elective surgery. Request sickle negative blood.

  The decision to transfuse pre-operatively depends on several factors including the pre-operative haemoglobin level compared with the steady state haemoglobin, the patient’s exercise tolerance, previous history and the type of surgery. Children with previous sickle problems (chest crisis, frequent painful crises) and obstructive sleep apnoea are at greatest risk. There is evidence that exchange transfusion confers no benefit pre-operatively over simple top up transfusion in minor or intermediate surgery. ‘No transfusion’ is safe for short procedures, but recent evidence suggests a benefit to simple top up transfusion even in ‘low risk’ surgery.

The following are a general guide but each patient must be discussed between the haematology team, surgeon and the anaesthetist.

4 **Transfusion Recommendations for Elective Surgery**

All cases of SCD patients must be brought to the attention of a Consultant Anaesthetist, Consultant Surgeon and Consultant Haematologist and a written plan formulated and put in their notes prior to their surgery. See M3/Form1/1096

- Group1, short procedure (eg grommet insertion), no extra risk factors
  - **ACTION**: discuss top up transfusion if Hb<90g/L
- Group 2, intermediate risk surgery such as tonsillectomy, adenoidectomy or cholecystectomy
  - **ACTION**: top up transfusion to 90-100g/L
- Group 3, major surgery – eg thoracotomy, or children who have had previous chest crises or severe vaso occlusive crises
  - **ACTION**: exchange transfusion or sequential top ups to achieve HbS level <30% and Hb <120g/L

In some cases at SC(NHS)FT surgery has been facilitated in children whose parents would not consent to transfusion by using a combination of oral hydroxycarbamide and s/c erythropoietin for 12 weeks prior to surgery. This is a consultant decision after careful discussion with the family.
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5. References


2. GOS Operative Management of Children with Sickle Cell Disease Guidelines


4. Use of short term hydroxycarbamide and once weekly erythropoietin to avoid transfusion in children with sickle cell anaemia undergoing Adenotonsillectomy. Welch JC. Poster presentation at British Society for Haematology April 2011

5. Howard et al. Pre-Operative Transfusion Reduces Serious Adverse Events in Patients with Sickle Cell Disease: Results from the Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS) Randomised Controlled Multicentre Clinical Trial. Oral and poster presentation 53rd American Society of Haematology Annual Meeting and Exposition December 2011

6. M3/Form1/1096 - Peri-Operative plan for Patients with Sickle Cell Disease