Pre Operative Assessment of Patients with Haemoglobinopathies

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Royal Liverpool University Hospital
Haemoglobinopathies

• Haemoglobinopathies are a group of recessively inherited genetic conditions affecting the haemoglobin component of blood.
• They are caused by a genetic change (mutation) in the haemoglobin. More than 1,000 mutations have been identified that result in either haemoglobin variants or thalassaemias.
The most significant haemoglobinopathies result in either:

• change in the structure and quality of the haemoglobin
• reduction in the quantity of haemoglobin produced.
Types of haemoglobinopathy - NB. all are genetically inherited

- **Qualitative – Sickle Cell Disease**
  - Affecting the structure and therefore maybe the function of the globin molecule
  - Eg. HbS, HbC, HbD Punjab, HbE, hundreds of others

- **Quantitative - Thalassaemias**
  - Affecting the amount of globin molecules produced
  - **Either** alpha chain defects – alpha thalassaemias
    - 4 genes, 2 from each parent,
  - **Or** beta chain defects – beta thalassaemias
    - 2 genes, 1 from each parent
Who is at risk?

Ethnic origin is critical!
Why is SCD important?

• It is the most common inherited condition in the World and in the UK
  – 15,000 affected in UK
  – 1- 2,000 affected in UK with Thalassaemia

• It is highly clinically significant causing lifelong morbidity and early mortality
Types of Sickle Cell Disease?

- Haemoglobin SS (Sickle Cell Anaemia)
- SS most prevalent and severe
- Haemoglobin SC
- Haemoglobin Sβ⁰ thalassemia
- Haemoglobin Sβ⁺ thalassemia
- Haemoglobin SD
- Haemoglobin SE
- Haemoglobin S Lepore
- Hemoglobin S/O(Arab)
Pathophysiology

Abnormal haemoglobin polymerises at low oxygen tension

Red cells become rigid

Haemolysis  Vaso-occlusion
Clinical Implications and Complications of Sickle Cell Disease

- Eye Problems
- Respiratory
  - Acute Chest Syndrome
  - Cardiomegaly
- Renal disease/Nephropathy
- Priapism
- Infection
- Pain
- Stroke
- Pulmonary hypertension
- Hepatic fibrosis, cirrhosis and cancer
- Hyposplenism
- Splenic sequestration
- Gallstones
- Avascular Necrosis
- Leg Ulcers
- Pain
Main Surgical Interventions

- Toncilectomy
- Cholycystectomy
- Splenectomy
- Joint replacements (Hips, Shoulders)
- Cardiac Surgery
Routine preoperative tests for elective surgery

NICE guideline
Published: 5 April 2016
nice.org.uk/guidance/ng45
1.4 Sickle cell disease or sickle cell trait tests

1.4.1  Do not routinely offer testing for sickle cell disease or sickle cell trait before surgery.

1.4.2  Ask the person having surgery if they or any member of their family have sickle cell disease.

1.4.3  If the person is known to have sickle cell disease and has their disease managed by a specialist sickle cell service, liaise with this team before surgery.
To Screen or not to Screen?

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 (GSTT).
Complications during Surgery in Sickle Cell Trait

- Isolated case reports of unusual adverse events raise the possibility that surgery involving hypoxia or reduced perfusion could result in vaso-occlusion and serious complications for people with sickle cell trait.

Hb 136, RCC 4.47, MCV 88.0, MCH 29.6
Sickle Cell Carrier (HbA + HbS)
No Evidence of Thalassaemia

Haemoglobin A2  n/a  2.0 – 3.5
Haemoglobin F  0.7 % 0.2 - 2.0
Haemoglobin S  35.2 %
Haemoglobin C  0.0 %
Haemoglobin D  0.0 %
Because surgery exposes patients to many of the factors that are known to precipitate red blood cell sickling, persons with SCD undergoing surgery require meticulous clinical care to prevent perioperative sickle cell–related complications.

Even with meticulous care, approximately 25% to 30% of patients will have a postoperative complications.

Triggers for Sickle Cell Crisis

• Hot and cold weather, variations in temperature
• Dehydration
• Stress
• De-oxygenation
• Infection
• Over exertion
• Low perfusion states
• 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.

• (NB This includes HbSS, HbSC, Hb SBthal, HbSE and HbSD)
<table>
<thead>
<tr>
<th>Haemoglobin</th>
<th>Value</th>
<th>Reference Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hb 100, RCC 2.30, MCV 122.4, MCH 43.4</td>
<td>Homozygous Sickle Cell Anaemia (HbSS)</td>
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<tr>
<td>Haemoglobin A2</td>
<td>2.0 % 2.0 - 3.5</td>
<td></td>
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<tr>
<td>Haemoglobin F</td>
<td>* 25.7 % 0.2 - 2.0</td>
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<tr>
<td>Haemoglobin S</td>
<td>64.6 %</td>
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<tr>
<td>Haemoglobin C</td>
<td>0.0 %</td>
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<tr>
<td>Haemoglobin D</td>
<td>0.0 %</td>
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<table>
<thead>
<tr>
<th>Haemoglobin</th>
<th>Value</th>
<th>Reference Range</th>
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</thead>
<tbody>
<tr>
<td>Hb 72, RCC 2.55, MCV 81.4, MCH 28.4</td>
<td>Homozygous Sickle Cell Anaemia (HbSS)</td>
<td></td>
</tr>
<tr>
<td>Haemoglobin A2</td>
<td>NA</td>
<td></td>
</tr>
<tr>
<td>Haemoglobin F</td>
<td>* 4.5 % 0.2 - 2.0</td>
<td></td>
</tr>
<tr>
<td>Haemoglobin S</td>
<td>83.3 %</td>
<td></td>
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<tr>
<td>Haemoglobin C</td>
<td>0.0 %</td>
<td></td>
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<tr>
<td>Haemoglobin D</td>
<td>0.0 %</td>
<td></td>
</tr>
<tr>
<td>Sample</td>
<td>Hb 126, RCC 4.69, MCV 76.6, MCH 26.8</td>
<td>Hb 127, RCC 5.56, <strong>MCV 67.7</strong>, MCH 22.8</td>
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<tr>
<td></td>
<td><strong>HbSC Disease (HbSC)</strong></td>
<td>Sickle Cell / Beta Thalassaemia (HbSBThal)</td>
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<tr>
<td></td>
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<td></td>
</tr>
<tr>
<td>Haemoglobin A2</td>
<td>NA</td>
<td>Haemoglobin A2</td>
</tr>
<tr>
<td>Haemoglobin F</td>
<td>0.9 % 0.2 - 2.0</td>
<td>Haemoglobin F</td>
</tr>
<tr>
<td>Haemoglobin S</td>
<td>46.0 %</td>
<td>Haemoglobin S</td>
</tr>
<tr>
<td>Haemoglobin C</td>
<td>44.0 %</td>
<td>Haemoglobin C</td>
</tr>
<tr>
<td>Haemoglobin D</td>
<td>0.0 %</td>
<td>Haemoglobin D</td>
</tr>
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Treatments

- Hydroxycarbamide
- Blood Transfusion
- Penicillin V
- Folic Acid
- Analgesia
Blood Transfusion - HbSS and HbSB$^0$Thal

All patients with HbSS and HbSB$^0$thalassaemia should be offered pre-operative transfusion 2-10 days prior to surgery.

Minor or moderate risk operations:
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 Haem team)

High risk operations: Exchange blood transfusion 2-10 days prior to surgery aiming for pre-op Hb of 100g/l and HbS% of <60%.
Blood Transfusion - HbSC/other genotypes

In view of the lack of evidence, decisions should be made on a case-by-case basis.

High risk procedure:
Offer exchange transfusion 2-10 days prior to surgery aiming for a preoperative Hb of 100g/L and HbS+C of <30%

Low or moderate risk procedure
If Hb <90g/L offer a top up transfusion aiming for a preoperative Hb of 100g/L
If Hb is >90g/L no transfusion unless very severe disease phenotype
Emergency Surgery

• Simple transfusion should be given preoperatively if Hb <90 g/l provided this will not result in undue delay to surgery. If transfusion is likely to cause an unacceptable delay to surgery, it is reasonable to proceed to surgery while arranging to transfuse the patient intra- or post-operatively if necessary.
Red Cell Selection

- As a minimum, red cells should be matched for Rh (D, C, c, E, e) and K antigens
- Red cells should be HbS negative

NSAHG, Ce, M, Le(a), Fy(a), aE, NSEA, e, CMV
Complications of Blood Transfusion

• Alloimmunisation
• Hyperhemolysis (DHTR)
# Sickle Cell Disease Perioperative Management Plan

<table>
<thead>
<tr>
<th>Name</th>
<th>Procedure</th>
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<tbody>
<tr>
<td>Hospital No</td>
<td>EVAR</td>
</tr>
<tr>
<td>NHS No</td>
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<tr>
<td>Address</td>
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**NOTE**

This Patient has Sickle Cell Disease. There is an increased risk of both Surgical (e.g., Infection, Thrombosis) and Sickle Cell Related (e.g., Acute Painful Crisis, Acute Chest Syndrome) complications in the perioperative period.

Unless in an emergency situation DO NOT Transfuse without discussion with Haemoglobinopathy Team.

Please discuss any concerns with a member of the Haemoglobinopathy Team.

<table>
<thead>
<tr>
<th>Date/Time</th>
<th>Plan</th>
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<tbody>
<tr>
<td>Wednesday</td>
<td>25/11/15 12:00 Admit to ward B4. Bloods as follows: FBC, Haemoglobin Profile, Group and Save, Cloting Screen + Fibrinogen, full profile. Review by Chest Physiotherapist to discuss Incentive Spirometry.</td>
</tr>
<tr>
<td>Thursday</td>
<td>26/11/15 To Interventional Theatre for insertion of apheresis line (7) to supply Bloods as follows: 2 x Group and Save (4 match 12 units for red cell exchange)</td>
</tr>
<tr>
<td>Friday</td>
<td>27/11/15 To Therapeutic Apheresis Unit (7x) for Red Cell Exchange Transfusion (usually takes 2-3 hours) Aim: Reduce HbS level to &lt;30% Increase Hb to 100g/L. Bloods: recheck Haemoglobin Profile post exchange</td>
</tr>
</tbody>
</table>

**NOTE**

Patient at increased risk of infection – if Temperature >38°C for clinical review and consideration of broad spectrum antibiotics

Patient at risk of Acute Chest Syndrome – if SpO2 fall below 94% (or >3% below baseline) for clinical review and discussion with Haemoglobinopathy Team

**Haemoglobinopathy Team Contacts**

Monday – Friday 09:00-17:00 Haematology SR for Coagulation via Switchboard

Andy Houghton (SCD Nurse Specialist) – RLH Sleep 49.1

Dr David Simcox (Consultant Haematologist) – via Switchboard

Out of Hours Contact Haematology SR on-call via RLH Switchboard
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
Post-operative management (1)

- 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.
- $O_2$ therapy for 48 hours to maintain oxygen saturations >94% (major cases)
- Consider ventilatory support with incentive spirometry or continuous positive airways pressure
Post-operative management (2)

- Effective analgesia
- 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.
Team Work

• Poor team work

• Good team work
• Thank You
Thalassaemia

- Patients should be carefully assessed pre-operatively with specific reference to complications relating to thalassaemia including cardiac, thrombotic, endocrine and metabolic disturbances.

- **There should be close liaison between the surgical, anaesthetic, and haematology teams in planning surgery, and shared care arrangements should be agreed prior to a surgical admission.**

- Patients undergoing urgent surgery should always be discussed with the Haematology team, during preparations for theatre.

- Patients should have been given adequate information regarding thalassaemia-specific and other issues related to surgery to allow informed consent.
Blood Transfusion

- Blood transfusion for the TDT patient should be planned around elective surgery, to allow surgery to be undertaken with optimal Hb level (100 – 120 g/l).
- Special blood requirements should be communicated well in advance to the surgical unit’s transfusion laboratory.
Team Work

• Poor team work
• Good team work