# Blood Transfusions in Children with Haemoglobinopathies

<table>
<thead>
<tr>
<th>Version:</th>
<th>2</th>
<th>Date:</th>
<th>22\textsuperscript{nd} April 2010</th>
</tr>
</thead>
<tbody>
<tr>
<td>Authors:</td>
<td>Dr S Height, Dr D Rees</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Responsible committee or Director:</td>
<td>Child health</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Review date:</td>
<td>23\textsuperscript{rd} April 2012</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Target audience:</td>
<td>Paediatricians, haematologists, paediatric surgeons, anaesthetists</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stakeholders/committees involved in guideline development:</td>
<td>Paediatricians, haematologists, transfusionists</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

---

<table>
<thead>
<tr>
<th>Ratified by:</th>
<th>Clinical Guidelines Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>Date ratified</td>
<td></td>
</tr>
<tr>
<td>Date when policy comes into effect:</td>
<td></td>
</tr>
</tbody>
</table>
Blood Transfusions in Children with Haemoglobinopathies

General Considerations

- These guidelines are to be read in conjunction with the Trust’s ‘Blood Transfusion Policy & Procedure - The prescription, collection and administration of blood components’ and ‘Guidelines for the use of Blood Components’ (both on Cliniweb).

- Blood transfusion is used for specific indications and may be either:
  a) Simple additive or “top-up” transfusion
  b) Exchange transfusion

- Children likely to receive frequent or regular blood transfusions should be vaccinated against Hepatitis B as early as possible if not already immune.

- Blood Bank should have a record of the extended phenotype of all children who attend clinic regularly.

- All new patients should have two 2-5mls EDTA sample sent to Blood Bank for:
  ABO and RhD typing
  Antibody screen
  Extended red cell phenotyping: (Rh, Kell, Fy, Jk and MNS
  S– s– patients should be typed for U).

- The cross match sample and request form must contain diagnosis and full patient identification - surname, first name, date of birth, gender, hospital number or A&E number.

- Request sickle negative blood.

- Request CMV negative blood if <1 year of age or CMV status unknown or negative.

- Calculate volume needed - see specific transfusion guideline (see below).

- Order the blood indicating when and where it will be needed.

- Blood Bank need 24 hours notice to order phenotyped blood for ‘routine’ transfusions.

- For emergency transfusions, blood can be ordered if blood bank has the patient’s phenotype. Contact Blood Bank to discuss as soon as the need for emergency transfusion has been identified - if blood has to be ordered from Tooting and then cross-matched there may be a delay of ~ 4-6 hours.

Simple additive or “top-up” transfusion in Sickle Cell Disease

Indications
• **Acute anaemia**
  - Splenic or hepatic sequestration
  - Aplasia due to Parvovirus infection
• Acute chest syndrome with a fall in haemoglobin of more than 1g/dl Hb
• Preoperatively
• Primary and secondary stroke prevention

**Aim**
- **To improve oxygen delivery/clinical condition**
- To restore Hb to normal steady state
- The target Hb should usually be less than 10 g/dl or haematocrit 0.35 since this is likely to cause an increase in blood viscosity.

**Volume of blood for “top-up” transfusion**
- \[\text{Desired Hb - Actual Hb} \times \text{Weight (kg)} \times 3 = \text{Volume of packed cells}\]

**Rate**
- The normal rate of red cell transfusion is around 5ml/kg/hour.
- Frusemide is not usually given with transfusions in SCD because of the increase in viscosity that may result.
Exchange Transfusion in Sickle Cell Disease

Indications

- Severe acute chest syndrome likely to require respiratory support
- Stroke (or TIA)
- Multiorgan failure
- Retinal artery occlusion, hepatic failure
- Priapism unresponsive to therapy

Aim

- To achieve an HbS% of <30%, final Hb \( \leq 11.5 \text{g/dl} \) and haematocrit of <0.35
- This is achieved by performing a total exchange of 1 – 2 times the calculated blood volume (70 x weight in kg)
- A manual exchange usually achieves this over 2 exchanges lasting 2 - 4 hours each.
- An automated exchange using a cell separator allows the exchange to be completed as a single procedure and may be possible for patients >30kg – please discuss on an individual basis.

Volume of packed cells (ml) for each exchange

- The exchange volume will depend on the starting haemoglobin level and HbS%. In particular, the child may have had a marked fall in haemoglobin and/or a simple top-up transfusion may have been already given
- Plan a total exchange of 1 – 2 times the calculated blood volume
- The volume of packed cells (in ml) \text{for each} exchange = 0.6 \times 70 \times \text{weight (kg)}
- This volume exchange would need to be repeated nearly 3 times to achieve the total exchange as indicated above, however, in practice it is not usually necessary to perform more than 2 procedures

Procedure

- Order the blood urgently and send sample for cross-match.
- The exchange will usually be performed on HDU or PICU and the patient should be discussed with the consultant on HDU/PICU.
- Depending on the size of the patient and their clinical condition, the exchange may require the siting of an arterial line.
- Request the following investigations prior to exchange:
  - Hb, PCV and platelets
  - INR, APTR and Fibrinogen
  - Renal, liver & bone profiles and glucose
- Access - see above.
  - Ideally two ports, one each for venesection and transfusion.
  - A single line with a three-way tap is an alternative for larger children.
- The exchange protocol for each patient should be individually calculated and will be determined by many factors including starting Hb/HbS%, target Hb/HbS%, clinical condition of the patient, venous/arterial access.
- The planned protocol should be recorded on a chart, such as that at the end of this guideline.
- Fluid balance should be maintained throughout the procedure as much as possible. An appropriate volume of Normal saline should be infused every hour to maintain isovolaemia.
- Due to differences in the haematocrit of transfused red cells and whole blood, every 4ml of blood venesected, is replaced by 3ml packed cells and 1ml N saline.
- Document the procedure accurately on a form such as that at the end of this guideline.
• At the end of the first exchange send samples for
  
  Hb, PCV and platelets
  INR, APTR and Fibrinogen
  Renal, liver, bone profiles and glucose
  and
  HbS% 

• Depending on the patient’s clinical state, discuss the need to proceed to a second exchange.
• Repeat the investigations after each exchange.
• Ensure that at the end of the exchange the final Hb \leq 11.5 and PCV \leq 0.35.
Regular Transfusion in Sickle Cell Disease and Thalassaemia

Indications

**Sickle Cell Disease**
- Primary and secondary stroke prevention
- Recurrent sickle chest syndrome when hydroxyurea has failed or inappropriate
- Frequent episodes of acute pain when hydroxyurea has failed or inappropriate

**Thalassaemia**
- Transfusion dependent thalassaemia

Aims
- Maintain trough haemoglobin between 9-10g/dl
- Maintain HbS (or HbS+HbC) below target – typically 30% or 50% (SCD only)

Procedure
- Admission to Philip Isaac’s Ward – discuss and book with staff nurses initially (extension 4200)
- Plan to give transfusion every 3-4 weeks as a ‘top-up’. The frequency and post-transfusion target haemoglobin are specified at the monthly MDT meetings, and recorded on EPR
- \( [\text{Target Hb} - \text{Actual Hb}] \times \text{Weight (kg)} \times 3 = \text{Volume of packed cells} \)
- Patients who may be candidates for BMT should receive CMV negative blood if they are CMV negative or status is unknown.
- Check Hepatitis B status and immunize.
- Check Hepatitis C antibodies and anti-HBsAb annually and give booster when indicated.
- Children on regular transfusion programme will need to start iron chelation after about 12 months of transfusion or once ferritin is consistently more than 1000μg/l (see separate guideline).
- Monitoring for complications of iron chelation toxicity and/or iron overload – (see separate guideline).
- Patients on chronic transfusion should be reviewed by medical staff at each visit.
- Patients on transfusion must be given clinic appointments for formal review every 3-4 months and stroke patients will be seen periodically in the joint neurology/haematology clinic if they have had a stroke.

Monitoring Patients on Regular Transfusion
- All SCD and thalassaemia patients will be discussed at the monthly haemoglobinopathy MDT Meeting.
- This meeting will monitor, trough haemoglobin levels, HbS%, serum ferritin, volume of transfused blood, vitamin D levels and vaccination status.
- Adjustments will me made at this meeting to achieve target Hb/HbS levels which may typically involve altering the frequency of transfusion and target haemoglobin.
- Adjustments are recorded in the notes and on EPR by the clinical nurse specialist, Sandra O’Driscoll.
- Annual review tests including asessement of endocrine, cardiac and hepatic function will be organized by the clinical nurse specialist and reviewed in the MDT meeting.

**Transfusion and surgery in SCD**
• Minor and straightforward procedures (e.g. tonsillectomy, appendicectomy, possibly cholecystectomy) can be safely undertaken without transfusion in most patients. This must be reviewed for individual patients, particularly those with a previous history of severe acute chest syndrome. Send sample to blood bank for G+S (and phenotyping if not previously performed)
• Transfusion should be performed preoperatively for major procedures; hip/knee replacement, organ transplantation, eye surgery, neurosurgery, cardiovascular surgery, major abdominal surgery.
• Transfusion should ideally be given a few days prior to surgery.
• Top-up transfusion to Hb 8 – 10g/dl is as effective as exchange transfusion and may be safer.
• Please discuss with consultant if unsure how to proceed.
• Patients for low and moderate risk surgery should be considered for entry into the TAPS trial of preoperative transfusion. Please discuss with one of the paediatric haematologists or clinical nurse specialist.

**Transfusion and surgery in Thalassaemia**
• Ideally, surgery should be performed 1-2 weeks after the last transfusion in children with thalassaemia major. This may mean adjusting the date of the planned transfusion to minimize the risk of any surgery.
Children of Jehovah’s witnesses

- Parents who are Jehovah’s Witnesses may not give consent for the use of cellular blood products or plasma for their child. However, sensitive discussion of the situation may avoid the need for legal intervention in order to treat the child. It is very important to explain to the older child what is happening and why transfusion is necessary since they too may be distressed by the evident conflict between their parents’ beliefs and the need for transfusion.

- Some parents in this situation may sign a statement acknowledging that in a life-threatening situation the doctors caring for the child will want to give blood and that they understand the medical staff carry the responsibility for that decision although they do not agree.

- The Jehovah’s Witness Society has been helpful in providing support for individual children undergoing transfusion, with practical suggestions such as covering the bag and giving set to minimize the anxiety it may cause.

- In the event of parents refusing to allow transfusion in a life-threatening situation, then legal advice must be sought to allow treatment to proceed in the interests of the child.

- In some situations, transfusion may be avoided by the use of recombinant EPO which is usually acceptable to Jehovah’s Witnesses.

- Discuss with a consultant.

These protocols were updated 04/10.

Dr David Rees
Dr Susan Height

April 2010
**Example of Form Used to Plan and Record Exchange Transfusion**

Exchange transfusion protocol for _________________

Take blood for urgent cross-match, FBC, haemoglobin electrophoresis, urea and electrolytes, LFTs, clotting, hepatitis A,B and C serology before exchange.

<table>
<thead>
<tr>
<th>Time</th>
<th>Venesected blood</th>
<th>Transfused packed cells</th>
<th>N saline infused</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-15 minutes</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>15-30 minutes</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>30-45 minutes</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>45-60 minutes</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>1h-1:30</td>
<td></td>
<td></td>
<td>x</td>
</tr>
<tr>
<td>1:30 -1:45</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>1:45-2:00</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>2:00-2:15</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>2:15-2:30</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>2:30-3:00</td>
<td></td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>3:00-3:15</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>3:15-3:30</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>3:30-3:45</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>3:45-4:00</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>4:00-4:30</td>
<td></td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>4:30-4:45</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>4:45-5:00</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>5:00-5:15</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>5:15-5:30</td>
<td>x</td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>5:30-6:00</td>
<td></td>
<td>x</td>
<td></td>
</tr>
<tr>
<td>Total volume</td>
<td>A+B</td>
<td>A</td>
<td>B</td>
</tr>
</tbody>
</table>

Check FBC, Hb electrophoresis, urea and electrolytes, LFTs and clotting 30 minutes after end of exchange.
If Hb>12g/dl – vencect to target Hb 10.5g/dl
If Hb<9.5g/dl – vencect to target Hb 10.5g/dl