

GSTFT Clinical Practice Guideline

**Use of Blood Transfusions in Children with Sickle Cell
Disease**

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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
 - c) Hyper-transfusion
- Organize Hepatitis B immunization 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 a 2-5mls EDTA sample sent to Blood Bank for:
 - ABO 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.
- For planned 'routine' transfusions – if previously transfused -

3–14 days ago	→ Send sample <24hrs before transfusion
14–28 days ago	→ Send sample <72hrs before transfusion
29 days–3 months ago	→ Send sample <1 week before transfusion
- 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 2422 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

- Splenic or hepatic sequestration
- *Aplastic crisis*
- Fall in Hb to < 5g dl
Consider top-up in child with chest symptoms with a fall in Hb (discuss with consultant)
- Preoperatively (see below)

Aim

- To restore Hb to normal steady state
- The Hb should never be raised acutely to >10 g/dl or haematocrit to >0.35 since this is likely to cause an increase in blood viscosity).

Volume of blood for “top-up” transfusion

- $[\text{Desired Hb} - \text{Actual Hb}] \times \text{Weight (kg)} \times 3 = \text{Volume of packed cells}$

Rate

- The normal rate of red cell transfusion is around 5ml/kg/hour.
- Frusemide is not given with transfusions in SCD because of the increase in viscosity that may result.

Exchange Transfusion in Sickle Cell Disease

Indications

- Acute chest syndrome.
- Stroke (or TIA)
- Severe infection (meningitis, pneumonia)
- Retinal artery occlusion, hepatic failure
- Priapism unresponsive to therapy

Aim

- To achieve an HbS% of <20%, final Hb \leq 12.5/haematocrit of <0.35
- This is achieved by performing a total exchange of 1.5 – 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 larger teenage patients – please discuss on an individual basis.

Volume of packed cells (ml) for each exchange

- Plan a total exchange of 1.5 – 2 times the calculated blood volume
- The volume of packed cells (in ml) **for each** exchange = $0.6 \times 70 \times \text{weight (kg)}$
- NB 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 (Blood Bank Ext 2422) and send sample for cross-match.
- Depending on the size of the patient and their clinical condition, the exchange may require the siting of an arterial line and a PICU bed for the procedure.
- Discuss with PICU as soon as the decision to exchange the patient is made.
- 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.
- At start of procedure –
 - If PCV <0.20 → transfuse packed cells (8ml/kg body weight) and then proceed.
 - If PCV >0.20 → consider infusing N/saline (10–20ml/kg body weight) and then proceed.
- Normal saline (not FFP or albumin) should be used as volume replacement at the beginning of the exchange prior to starting venesection to avoid dropping the circulating blood volume.
- Maintain isovolaemia by withdrawing the patient's blood and replacing with donor blood at the same rate.
- Document the procedure using the exchange transfusion proforma
- 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 ≤ 12.5 and PCV ≤ 0.35 .

Hyper-transfusion in Sickle Cell Disease

Indications

- CVA – to prevent recurrence
- To prevent the development of stroke in children with sickle cell disease with Doppler and/or MRI evidence of cerebrovascular infarction/haemorrhage in the absence of clinical evidence of stroke.
- Recurrent sickle chest syndrome (consider Hydroxyurea)
- >3 severe vaso-occlusive crises/year (consider Hydroxyurea)

Aims

- Maintain Hb between 10.0 and 14.5g/dl
- Maintain HbS below 30% (or HbS + HbC below 30%)

- In some patients, a less intensive regimen maintaining the HbS below 50% may be sufficient for stroke prevention after 3 years – discuss with consultant.

Procedure

- Admission to Mountain Ward – discuss and book both Drs and Nurses' diaries
- Plan to give transfusion every 4 weeks as a 'top-up':

$$[\text{Desired Hb} - \text{Actual Hb}] \times \text{Weight (kg)} \times 3 = \text{Volume of packed cells}$$

- Patients who may be candidates for BMT should receive CMV negative blood if they are CMV negative or status is unknown. **NB all blood are leuco-depleted and for practical purposes CMV free**
- Check Hepatitis status and immunize those negative for both antigen and IgG antibody level <100 U/L
- Check Hepatitis C antibodies and anti-HBsAb annually and give booster when indicated.
- Children on hyper-transfusion programme will need to start s/c desferrioxamine infusion once ferritin >1000ng/ml (see separate guideline).
- Monitoring for complications of desferrioxamine toxicity and/or iron overload – (see separate guideline).
- Patients on chronic transfusion should be reviewed by medical staff and complete information sheet 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.

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 chest syndrome – please discuss.
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, and considered for 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.

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 consultant on call.