NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Adult Guideline

Red Cell Exchange Transfusion in Sickle Cell disease

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

Review: December 2027
NSD610-017.12 V4

Introduction

Most patients with sickle cell anaemia are relatively asymptomatic despite baseline Hb concentrations between 50-120g/l as HbS is a low-affinity haemoglobin and oxygen delivery to tissues is enhanced. **Chronic steady state anaemia alone is not an indication for transfusion.** Top-up transfusion increases whole blood viscosity and may aggravate sickling. Top up transfusion is not indicated for uncomplicated sickle crises (see separate guideline on perioperative management).

Exchange transfusion is a potentially life-saving procedure that allows correction of anaemia *without* increasing blood viscosity and may improve tissue oxygenation whilst reducing microvascular sickling.

The aim of exchange transfusion is to lower the HbS level to 30% or less while keeping the Haemoglobin close to 100g/dl. C*linical benefit may be seen even with a partial manual exchange*.

Prior to embarking on an exchange procedure, the case must be discussed with the Consultant Haematologist on call.

Indications for Exchange Transfusion

- **Acute cerebrovascular event**
- **Acute chest syndrome**
- **Multi-organ failure**
- **Preoperatively in selected cases**

Automated red cell exchange is preferred being quicker and much more effective at reducing HbS percentage and should be the standard of care for elective procedures. However, if emergency exchange is indicated and automated exchange cannot be performed in a timely manner then manual exchange should not be delayed. Even a partial manual exchange may have great clinical benefit whilst awaiting a definitive procedure.

Manual Red Cell Exchange Transfusion

1. Background

This procedure should only be performed by suitably qualified staff under the supervision of the responsible Consultant Haematologist.

Staff should be familiar with the procedure and management of associated complications.

Adult patients may require insertion of a femoral or internal jugular line but those with good peripheral access can be managed with one or preferably two large bore IV cannulae.

Trust policies for safe blood transfusion must be always adhered to.

2. Patient work up

Consent

Ensure the patient is consented for the exchange procedure. Often verbal consent alone will be practical when the patient is seriously ill.

Aspects which should be specifically mentioned include:

- access (peripheral vs vascath)
- Vasovagal episodes
- blood transfusion reactions and alloimmunisation
- transfusion related infection

Baseline Blood tests

- full blood count
- HbS percentage
- Urea and electrolytes
- Calcium
- Magnesium
- Liver function tests
- Virology – HIV, hepatitis B and C
- Ferritin levels, glucose, thyroid and endocrine function if appropriate (not relevant in emergency setting)

Cross match 6-8 units of packed red cells (depending on size of patient). These should be:

- Sickle negative
- Phenotypically matched (Rh and Kell as a minimum),
- if there are clinically significant red cell antibodies (current or historical) then the red cells selected should be negative for the corresponding antigens
- ideally less than 7 days old (not essential in an emergency)
- patients should have full red cell phenotype performed if not already known

N.B. It may be several hours before compatible blood is available for patients with allo-antibodies

Patient Assessment
Prior to the procedure, check:

- baseline observations (blood pressure, heart rate, temperature and oxygen saturations)
- patient weight (estimate in an emergency when accurate measurement not feasible).
- ensure adequate venous access
- check patient blood transfusion record for alloantibodies, and with patient directly to see if any history of transfusion related reaction or haemolysis

3. Equipment and supplies

- Baxter infusion pump
- blood giving set
- blood warmer
- 500mls normal saline (several packs)
- packed red cells (amount dependant on patient's size and condition, usually 6-8 units)
- Dynamap and tympanic thermometer
- Calcium gluconate in case of hypocalcaemia or hyperkalaemia

Equipment for Peripheral Access/Venesection

- Sterile gloves
- 16g Kimal access needle x1
- 18g or 20g cannula
- Chloraprep/skin preparation
- 4x10ml syringes
- 20mls of normal saline for flush
- Tegaderm dressing to secure cannula
- Mepore tape to secure Kimal needle
- Gauze swabs
- consent form (if applicable)
- blood collection form, observation chart and nursing documentation
- large sharps box
- weighing scales (to assist calculation of volume venesected)
- 3-way tap (useful in patients with limited peripheral access)
- sterile bungs (to allow repeated access of large bore venesection pack needle into 3 way tap extension set)
- Venesection packs

Patients requiring a central line
Follow local line insertion protocol

4. Method

Perform the venepuncture for the access (venesection) line and return lines. Ensure access on both sides is secured and both lines are flushed with normal saline prior to commencing the procedure.

Set up the giving set for the return line so that fluid/blood administered is warmed in the blood warmer.

Do not start until compatible blood is available on the ward

1. set up a bag of normal saline and run 500mls over 15 to 30 minutes to ensure the patient is adequately pre-hydrated (reduce rate/and or volume if concern over fluid overload or cardiovascular compromise)
2. ensure the blood is warmed prior to infusing
3. to venesect: remove 450-500mls of blood over 15-30minutes.
4. ensure local transfusion policies are adhered to, and documentation completed.
5. calculate the amount to be exchanged, dependant on the starting haemoglobin, as follows:
    - Hb >80g/l 5-8 units
    - Hb 60-79g/l 4-6 units
    - Hb <60g/l up to 4 units

Exchange Procedure if starting Hb >80g/l

Venesect 1st unit	***whilst***	replacing with 500mls normal saline stat
Venesect 2ndunit	***then***	transfuse 1st unit over 30-40 minutes
Venesect 3rd unit	***then***	transfuse 2nd unit over 1 hour
Venesect 4th unit	***then***	transfuse 3rd unit over 2 hours

Re-check FBC, HbS and electrolytes at this stage

If repeat Hb<90g/l	transfuse 4th unit and consider 5th unit (3hrs each)
If repeat Hb>90g/l	restart from “venesect 1st unit”

N.B. By removing **two** units of blood before transfusing the 1st unit, this method results in more efficient lowering of the HbS %. However, if the patient is cardio vascularly unstable, or becomes hypotensive during the venesection, the replacement transfusion should be started sooner, i.e. after venesection of the 1st unit.

If starting Hb 60-79g/l

Venesect 1st unit ***whilst*** transfusing 1st unit

Venesect 2ndunit ***then*** transfuse 2nd unit over 1hr and 3rd and 4th units over 3hrs

Further exchange may be required (see "Hb 8g/l") if insufficient clinical improvement/impact on HbS level

If starting Hb<60g/l

Top up transfusion to 80-100g/l (rate depending on clinical condition and baseline Hb) initially, discuss with Consultant Haematologist.

Formal exchange may be required (see "Hb 80g/l") if insufficient clinical improvement/impact on HbS%

5. Post procedure

Remove Kimal needle but leave cannula in, flush with normal saline.

Monitor vital signs at 15- and 30-minutes post procedure

Take bloods 30 minutes post procedure for:

- Full blood count
- HbS percentage
- Electrolytes, calcium and magnesium
- Coagulation screen and fibrinogen

Avoid final Hb of >100g/l or >10-20g/l above steady state Hb, particularly if the post-transfusion HS% exceeds 30% (risk of hyper viscosity) or <70g/l.

Watch for development of **hyperkalaemia** and **hypocalcaemia** during the exchange.

Ensure all transfusion documentation is completed correctly and returned to blood transfusion as per local policy.