# Manual red cell exchange in patients with Sickle Cell Disease (SCD)

## Introduction

Most patients with sickle cell disease 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 (**however see separate guideline on perioperative management.**

Exchange transfusion is a potentially lifesaving 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/l. Clinical 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. (Consider discussion with OUH Haematologist on call **and refer to the Unwell patient discussion check list** , http://nssg.oxford-haematology.org.uk/red-cell/documents/acute-management-sickle-cell-disease/S62-unwell-patient-discusion-checklist.pdf

## Indications for manual red cell exchange transfusions

**THIS IS AN EMERGENCY PROCEDURE ONLY**

*Manual exchange is only to be used in the acute setting where an automated service is unavoidably delayed. Note that the NHSBT provides a 24 hour service for automated exchange including an outreach service – automated exchange is always preferable.*

See Acute automated red cell exchange protocol: http://nssg.oxford-haematology.org.uk/red-cell/documents/transfusion-and-exchange/S5-1-acute-outreach-automated-red-cell-exchange-pathway.pdf

**Indications for red cell exchange:**

| Recommended | Should be considered |
|---|---|
| • Acute chest syndrome: severe clinical features or evidence of progression despite initial simple transfusion<br>• Acute ischaemic stroke<br>• Acute hepatic or splenic sequestration<br>• Emergency surgery (individual considerations) | • Acute multi-organ failure<br>• Mesenteric/girdle syndrome<br>• Severe sepsis<br>• Acute priapism<br>• Non-resolving acute painful crisis<br>• Acute intrahepatic cholestasis |

## Background

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

- Staff should be familiar with this procedural document and management of associated complications
- 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 and patient monitoring must be adhered to at all times
- All patients with haemoglobin disorders should receive only blood that has been subject to extended phenotyping: Blood Bank should be alerted that the patient has sickle cell disease prior to cross-matching

## Pre-procedure

### Consent

Where possible ensure the patient has given verbal consent for the exchange procedure as per standard transfusion practice.

Aspects that should be specifically mentioned include:

- Access (peripheral vs central)
- Vasovagal episodes
- Blood transfusion reactions and allo-immunisation
- Transfusion related infections

### Baseline Blood tests

- FBC, reticulocyte count
- Baseline coagulation screen
- HbS percentage (do not wait for the result)
- Urea and electrolytes, Calcium, Magnesium
- Liver function tests, LDH

- Virology – Hepatitis B (core antibody and surface antigen), Hepatitis C, HIV
- *Cross match 6-8 units of packed red cells (depending on size of patient). The units should be:

    - Sickle trait negative
    - Phenotypically matched (Rh and Kell as minimum)
    - Ideally less than 5 days old (not essential in an emergency)

NB patients should have full red cell phenotype performed if this is not already known and the patient has not had a recent transfusion

*• Hb >80g/l 5-8 units
- Hb 60-70g/l 4-6 units
- Hb <60g/l up to 4 units

***Urgently**, inform Transfusion Laboratory of planned manual exchange and check for history of previous transfusion reactions or allo-antibodies. The transfusion laboratory must be made aware of degree of urgency of exchange.

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

**Patient Assessment**

Prior to the procedure, check:

- Venous access
- Blood pressure: arms and leg, heart rate, respiratory rate, oxygen saturations, temperature
- Weight (or estimate)
- Connect patient to cardiac monitor, for continuous monitoring, including oxygen saturations

**Equipment and supplies**

- Infusion pump
- Blood giving set
- Blood warmer-if available
- 500ml 0.9% sodium chloride (several packs)
- Packed red cells (amount dependent on patient's size and condition, usually 6 - 8 units)
- Calcium gluconate in case of hypocalcaemia or hyperkalaemia
- Cardiac monitor
- Fluid balance chart/documentation

## Equipment for Peripheral Access/Venesection

- Sterile gloves
- 16g cannulae x 2
- Chloraprep/skin preparation
- 4x10ml syringes for saline flushes
- 50ml syringes for venesection if needed
- 20mls of normal saline for flush
- Dressings and tape to secure cannulae
- Gauze swabs
- Large sharps box
- Weighing scales (to assist calculation of volume venesected) if available
- 3 way tap (if only 1 access cannula can be achieved)
- Sterile bungs (to allow repeated access of large bore venesection pack needle into 3 way tap extension set)
- Venesection packs (if available)

## Patients requiring a central line

Follow local line insertion protocol

## Method

## Do not start until compatible blood is available on the ward

- Establish access for venesection and return lines
- Connect the three way tap and sterile bungs if needed
- Ensure access on both sides is well secured and both lines are flushed with normal saline prior to commencing the procedure
- Set up the blood giving set for the return line so that fluid/blood can be administered. Warmed if possible using a blood warmer
- To venesect, remove 450-500ml of blood over approximately 15-30 min into the venesection bag

  OR

  Blood can be aspirated from the line using 50ml syringes, which can be counted and discarded
- The procedure should be performed more slowly than described in patients with significant renal or cardiac abnormalities, or if cardiovascularly unstable
- The patient should be kept in overall fluid balance throughout the procedure. This may require the infusion of additional saline if small units of blood are provided.

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. Use a blood warmer if available.
3. To venesect: remove 450-500mls of blood over 15-30 minutes
4. Ensure local transfusion policies are adhered to, appropriate monitoring undertaken and documentation completed
5. Calculate the amount to be exchanged, dependent on the starting haemoglobin, as follows:
   - Hb >80g/l 5-8 units
   - Hb 60-70g/l 4-6 units
   - Hb <60g/l up to 4 units

**Exchange Procedure**

**If starting Hb >80g/dl**

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

Re-check FBC, coagulation screen, HbS and electrolytes at this stage and review clinical condition

- Discuss the repeat results with the Consultant Haematologist to decide whether further exchange should continue.
- Consider the need for plasma products to correct coagulopathy
- 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 cardiovascularly 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 2nd unit ***then*** Transfuse 2nd unit over 1hr and 3rd and 4th units over 3hrs
- Further exchange may be required (see "Hb 80g/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%

**Post procedure**

- Remove access needle but leave cannula in, flush with normal saline.
- Monitor vital signs at 15 and 30 minutes post procedure and then as clinically indicated
- Take bloods 30 minutes post procedure for:
  - FBC
  - HbS percentage
  - Electrolytes, calcium and magnesium
  - Coagulation

**Avoid final Hb of >110g/l (risk of hyperviscosity) or <70g/l**

- Watch for development of hyperkalaemia and hypocalcaemia during the exchange.
- Ensure all transfusion documentation is completed correctly and unused blood returned to blood transfusion as per local policy.

**Documentation and acknowledgements:**

Standards for the Clinical Care of Adults with Sickle Cell disease in the UK © Sickle Cell Society 2018

NHLBI (2014) Evidence- Based Management of Sickle Cell Disease, NIH.

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**Review**

| Name | Revision | Date | Version | Review date |
|---|---|---|---|---|
| Dr Wale Atoyebi | Pre-peer review | Jan 2013 | 1.0 | Jan 2015 |
| Dr Deborah Hay | Routine review | Aug 2015 | 1.2 | Jan 2017 |
| Dr Asif Khan | Routine review, network relevance | May 2017 | 2.0 | May 2019 |
| Dr Magbor Akanni | Routine review, HCC | Dec 2020 | 2.1 | Dec 2022 |
| Dr Wale Atoyebi | Inclusion of Unwell patient checklist and update of the baselines blood tests | October 2022 | 2.2 | October 2024 |