Automated red cell exchange transfusion in patients with sickle cell disease: Clinical guideline (HN-504b)

Background
Anaemia in sickle cell disease is often well tolerated, as HbS is a low affinity haemoglobin and oxygen delivery to tissues by HbS is enhanced relative to HbA. This, plus the observation that top-up transfusion increases whole blood viscosity and may aggravate sickling, means that chronic steady state anaemia alone is not an indication for transfusion and top-up transfusion is not indicated for uncomplicated vaso-occlusive episodes.

Automated red cell exchange transfusion (a-RCE) is a potentially life saving form of transfusion that allows correction of anaemia without increasing blood viscosity and it may therefore improve tissue oxygenation whilst reducing microvascular sickling. The aim of a-RCE is to raise the HbA level to 70% or more, while keeping Hb ~100 g/L. Clinical benefit may be seen even with a partial manual RCE, in urgent situations where automated RCE is not possible. See separate policy.

Prior to embarking on any acute red cell exchange, the case must be discussed with the Consultant Haematologist on call. Regionally, a-RCE is undertaken by NHSBT therapeutic apheresis service (Oxford), who offer both an emergency (24/7) and elective service.

Indications for automated red cell exchange (a-RCE)
The evidence base for a-RCE in terms of controlled clinical trials is limited; however, in the following clinical situations it is now accepted as best practice:

Acute
- Acute chest syndrome: severe clinical features or evidence of progression despite initial simple transfusion
- Acute ischaemic stroke
- Acute multi-organ failure
- Acute hepatic or splenic sequestration
- Mesenteric/girdle syndrome
- Acute intrahepatic cholestasis
- Severe sepsis
- Emergency surgery (individual considerations)

Elective
- Primary stroke prevention
- Secondary stroke prevention
- Vaso-occlusive episodes in pregnancy
- Current or previous complications in pregnancy
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- Repeated severe vaso-occlusive episodes, where hydroxycarbamide is contraindicated or ineffective
- Pulmonary hypertension
- Leg ulcers

**Acute a-RCE Referral/Transfer Pathway**

1. **Mon-Fri 9-5:** OUH haematology registrar (bleep 6888) via switchboard, 0300 304 7777.
   Haematology registrar will discuss indication with OUH Haemoglobinopathy consultant (Atoyebi, Hay, Roy) via switch board 0300 304 7777. In the case of regional referrals, also with the attending ward consultant (Lymphoid team) and ward establish bed availability for urgent transfer.

2. **Out of hours:** On-call OUH haematology registrar via switchboard 0300 304 7777 who will discuss indication with on call OUH Haematology consultant. Bed availability must be established.

3. Acute regional transfers MUST NOT be sent until a bed is identified. Patients MUST NOT be sent to the emergency department.

4. **OUH haematology refer to:**

5. **NHSBT Therapeutic Apheresis Service (TAS):** Medical referral.
   **Mon-Fri 0830-1630:**
   01865 387938 or 01865 234344
   **Out of hours: Mon-Fri 1630-0830 & 24h Weekends:**
   Contact NHSBT Oxford Hospital Services:
   01865 387963 or 01179 594666 (NHSBT On Call Consultant).

6. TAS will require patient’s height, weight, current HCT and HbS% if available, to calculate required blood volume.

7. Complete NHSBT referral [http://hospital.blood.co.uk/media/27366/frm5121-2.doc](http://hospital.blood.co.uk/media/27366/frm5121-2.doc) and email. Document link can also be found on NSSG/Adult haemoglobinopathies/NHSBT.

8. Notify blood bank as soon as indication and or transfer is agreed.

9. A group and screen sample should be sent to the JR blood bank and for extended red cell phenotyping if not previously undertaken. The volume of blood required for a-RCE is calculated by TAS staff in liaison with blood bank.

**Pre a-RCE baseline bloods**

- FBC, Reticulocyte %
- HbS % (do not wait for result in the acute setting)
- Clotting screen
Automated red cell exchange

Authorised by: Wale Atoyebi

• Urea and electrolytes
• Calcium
• Magnesium
• Liver function tests (Bilirubin, ALT, alkaline phosphatase, albumin)
• Viral serology – HIV, hepatitis B and C
• Also consider ferritin, glucose, thyroid and endocrine function if appropriate.
• Plus those required for a-RCE indications

**Post a-RCE bloods**

• FBC
• HbS%

**Agreed settings for acute a-RCE**

• Clinical Haematology Ward at the Churchill Hospital
• ICU at the Churchill or John Radcliffe Hospitals
• Women’s hospital observation area

**Venous access**

• Veins should be large enough to easily site a 16 G venflon in each antecubital fossa.
• Where peripheral venous access is inadequate, a suitable central apheresis line (renal vascular catheter or femoral line {available from DTU}). 9-5 the line insertion service should be used. Out of hours contact ICU.

**Patient information and consent**

The rationale for the procedure is to be explained to the patient and family by the haematology clinical team. NHSBT staff will explain the procedure itself and request written consent from the patient. A patient information leaflet is available on the NSSG or via [https://nhsbtdbe.blob.core.windows.net/umbraco-assets-corp/1949/red-cell-exchange-procedure.pdf](https://nhsbtdbe.blob.core.windows.net/umbraco-assets-corp/1949/red-cell-exchange-procedure.pdf)

**Nursing responsibilities**

NHSBT TAS are responsible for patient monitoring during the a-RCE, in relation to the transfusion and not the patients acute clinical state. The ward nursing team are responsible for the ongoing acute patient monitoring. NHSBT staff will complete all observations in SEND and use the joint approach nursing care plan (N120) [http://nssg.oxford-haematology.org.uk/oxford/clinical-care/N-120-joint-apheresis-dialysis-patient-line-management.pdf](http://nssg.oxford-haematology.org.uk/oxford/clinical-care/N-120-joint-apheresis-dialysis-patient-line-management.pdf)

**Post a-RCE monitoring**

Monitoring of vital signs (Temperature, pulse, BP, respiratory rate, oxygen saturations) should be carried out

• every 15mins for the first hour
• every 30 minutes for the next hour
• then as clinically indicated but a minimum of 4 hourly
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Additional monitoring will be needed depending on the indication for the a-RCE. In all cases, be alert for signs of delayed red cell transfusion reactions.

**Elective a-RCE referral**

Referral pathways for a-RCE
- Via Haemoglobinopathy MDT discussion
- Referral to Oxford haemoglobinopathy team (Dr’s Atoyebi, Hay, Roy) via orh-tr.clinicalhaematology@nhs.net
- Review at the Specialist Haemoglobinopathy Outreach clinic for discussion and coordination of red cell exchange.

Referral to NHSBT TAS
- Direct from local haematologist where Haemoglobinopathy MDT discussion has taken place
- By OUH if patient is seen in Outreach or Oxford clinic
- Complete NHSBT referral [http://hospital.blood.co.uk/media/27366/frm5121-2.doc](http://hospital.blood.co.uk/media/27366/frm5121-2.doc) and email.
- An individualised plan will be made regarding timing and location of the exchange and the requirement for central venous access.

Location of elective exchanges
- NHSBT TAS suite, Oxford
- Or Haematology Day Treatment Unit where patient condition or complex line insertion is required
- Or Women’s hospital observation area

**Ongoing management of red cell exchange programmes**

The frequency of exchanges is determined in conjunction with the OUH consultant team and is reviewed with NHSBT at the clinical service/TAS quarterly MDT.

Coordination of planned programmes:
- NHSBT are responsible for the coordination of the exchange procedures undertaken in the apheresis suite and women’s centre

References:

Adult Haemoglobinopathy Service


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<td>Dr Wale Atoyebi</td>
<td>Pre-peer review</td>
<td>Jan 2013</td>
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<td>Dr Deborah Hay</td>
<td>Routine review</td>
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<td>Dr Wale Atoyebi, Sandy Hayes, Specialist Nurse</td>
<td>Full review, updated references and pathways</td>
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