Clinical guideline: automated exchange blood transfusion in patients with sickle cell disease

HN-504b

Background
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. Top-up transfusion increases whole blood viscosity and may aggravate sickling. Therefore, chronic steady state anaemia alone is not an indication for transfusion and top-up transfusion is not indicated for uncomplicated sickle crises.

Exchange blood transfusion (EBT) 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 Hb ~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. In this Trust, automated EBT is undertaken by NHSBT based on the John Radcliffe Hospital site.

Indications for red cell exchange transfusions
The evidence base for blood transfusion in terms of controlled clinical trials is limited; however, there are certain clinical situations in which it is now accepted as best practice:

Acute indications
- Acute stroke
- Acute chest syndrome
- Severe sepsis
- Acute hepatic or splenic sequestration
- Multi-organ failure
- Progressive intrahepatic cholestasis

Elective Indications
- Primary stroke prevention
- Secondary stroke prevention
- Elective surgery
- Painful crises in pregnancy
- Current or previous complications in pregnancy
- Repeated severe painful crises
- Pulmonary hypertension
- Leg ulcers

Referral procedure for acute indications
- The final decision to carry out a red cell exchange transfusion will be made by a haematology consultant.
- This then needs to be discussed with the Consultant covering the NHSBT for the day (or contact the Apherisis unit on 01865 (2)20303)
- The NBS should be alerted as soon as the possibility of acute EBT is raised, and before 5pm where possible (even if the procedure is subsequently not required)
- Referral forms for EBT may be found on the TSSG Haematology website, under Transfusion/non-malignant haematology - http://clsmac70.ndcls.ox.ac.uk/tssg-haematology/transfusion/transfusion.html
  The patient’s height and weight must be recorded.
- A Group and Cross-match sample should be sent to the JR blood bank for extended red cell phenotyping. The volume of blood required for the transfusion is calculated by the NHSBT (Apherisis) staff in liaison with blood bank.

Referral procedure for elective red cell exchange transfusion
- A referral needs to be made to both the Consultant in charge of the Haemoglobinopathy service (Dr Wale Atoyebi) and Dr Sylvia Benjamin (NHSBT Consultant). The decision to embark on elective exchange will follow review by one/both consultants.
- An individualised plan will then be made regarding timing and location of the exchange and the requirement for central venous access. Formal referral to the NBS is via the proforma on the Haematology TSSG website http://clsmac70.ndcls.ox.ac.uk/tssg-haematology/transfusion/transfusion.html

Baseline blood tests prior to exchange transfusion
• FBC, Retics
• HbS %
• Urea and electrolytes
• Calcium
• Magnesium
• Liver function tests (Bilirubin, ALT, Alkaline phosphatase, albumin)
• Virology – HIV, hepatitis B and C
• Also consider ferritin, glucose, thyroid and endocrine function if appropriate.

Preparing the patient for exchange blood transfusion
• The patient must have adequate venous access for the procedure. The veins should be large enough to easily site a 16g venflon in the antecubital fossa.
• If the veins are inadequate, then a suitable apheresis line must be placed and functioning before the procedure (a renal vascular catheter is usually ideal).
• The rationale for the procedure needs to be explained to the patient. The NHSBT staff will explain the procedure itself and request formal consent from the patient. An information leaflet written for patients and their families is available.

Location of the exchange
Acute exchanges will take place in the following locations:
• Clinical Haematology Ward at the Churchill Hospital
• HDU/ITU at the Churchill or John Radcliffe Hospitals
Elective exchanges will take place at the Apheresis suite in the NHSBT, Oxford

Following a red cell exchange transfusion
Monitoring of vital signs (pulse, BP, oxygen saturations) should be carried out
• every 15mins for the first hour
• every 30 minutes for the next hour
• every 2 hours if they remain stable
Additional monitoring will be needed depending on the indication for exchange transfusion. In all cases, be alert for signs of delayed red cell transfusion reactions.

In all cases, a post-exchange HbS% should be checked.

Key contacts
Referrals for Exchange Blood Transfusion:
Dr Sylvia Benjamin  
Oxford STS Lead Consultant  
Oxford Therapeutic Apheresis Unit  
NHS Blood and Transplant, John Radcliffe Hospital, Oxford, OX3 9BQ  
Tel: 01865 220303  
Fax: 01865 38795  
sylvia.benjamin@nhsbt.nhs.uk

Ms Sophie Clarke  
Oxford STS Lead Nurse  
Oxford Therapeutic Apheresis Unit  
NHS Blood and Transplant, John Radcliffe Hospital, Oxford, OX3 9BQ  
Tel: 01865 220303  
Fax: 01865 38795  
sophie.clarke@nhsbt.nhs.uk

Urgent contacts for Oxford Specialist Therapeutic Apheresis Services (STS)  
During office hours (9am -5pm):  
• phone Apheresis Unit direct on 01865 (2)20303, or  
• phone Transfusion Registrar direct on 07764 280706, or  
• contact apheresis consultant of the day via the JR switchboard

Out of hours: phone NHSBT Oxford Hospital Services on 01865 387963, and ask to contact the Oxford consultant on the rota for apheresis (STS).

Documentation and acknowledgements:  
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Draft expert panel report on the management of Sickle Cell Disease, NHLBI, Bethesda, Maryland, USA, August 3 2012

Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK © Sickle Cell Society 2008

Authors:  
Dr Wale Atoyebi, Clinical Lead for Haemoglobinopathies

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<th>Date</th>
<th>Version</th>
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