# Sickle Cell Disease - Exchange Transfusion

<table>
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<tr>
<th>Title of Guideline</th>
<th>Guideline for Exchange Transfusions in Children and Young People with Sickle Cell Disease</th>
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Paediatric Haematology |
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| Guideline Number: | Version 2 |
| Explicit definition of patient group to which it applies | Children and young people under the age of 18 years with sickle cell disease |
| Abstract | This guideline describes the indications and procedure for performing an exchange transfusion in children and adolescents with sickle cell disease |
| Key Words | Paediatric, Children, Sickle cell disease, Exchange transfusion |

**Statement of the evidence base of the guideline – has the guideline been peer reviewed by colleagues?**

- 1a meta analysis of randomised controlled trials
- 2a at least one well-designed controlled study without randomisation
- 2b at least one other type of well-designed quasi-experimental study
- 3 well–designed non-experimental descriptive studies (ie comparative / correlation and case studies)
- 4 expert committee reports or opinions and / or clinical experiences of respected authorities
- 5 recommended best practise based on the clinical experience of the guideline developer

Consultation Process: Paediatric Haematology team

Target audience: Healthcare professionals caring for children and young people with sickle cell disease

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This guideline has been registered with the trust. However, clinical guidelines are guidelines only. The interpretation and application of clinical guidelines will remain the responsibility of the individual clinician. If in doubt contact a senior colleague or expert. Caution is advised when using guidelines after the review date.
Document Control

Document Amendment Record

<table>
<thead>
<tr>
<th>Version</th>
<th>Issue Date</th>
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<tbody>
<tr>
<td>V1</td>
<td>Sept 2011</td>
<td>Dr Simone Stokley Consultant Paediatric Haematologist</td>
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<tr>
<td>V2</td>
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<td>Dr Eleanor Jesky Consultant Paediatric Haematologist</td>
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Changes for new version:

- No significant changes required

Statement of Compliance with Child Health Guidelines SOP
This guideline has had only minor changes made and therefore this version has not been circulated to all for review. A previous version had been approved by circulation to senior team members.
Maria Moran, Clinical Guideline Lead
10th January 2019
Introduction

Red blood cell (RBC) transfusions are an integral part of the management of sickle cell disease. Transfusion of normal (non-sickle) blood into patients with sickle cell disease increases haematocrit (Hct) and simultaneously (by dilution) lowers the fraction of cells that contain HbS. By increasing the Hct, transfusion may also reduce the erythropoietic drive and decrease production of sickle haemoglobin. An exchange transfusion can be used to “replace” HbS by HbA. The end result of transfusion by any method is some combination of an increased haematocrit and a decreased proportion of RBCs that contain HbS.

Increases in Hct, although initially beneficial in improving oxygen delivery, are eventually counterbalanced by an opposing detrimental increase in blood viscosity. Using exchange transfusion it is possible to reduce the percentage of HbS without a detrimental rise in blood viscosity and without a significant change in blood volume.

It is more costly and labour intensive to perform an exchange transfusion than a simple top-up transfusion. An exchange transfusion also exposes the patient to a higher risk of transfusion complications (infections, alloimmunisation, need for central venous lines, etc.).

RBC exchange transfusion offers potential advantages over simple transfusion for the management of certain complications of sickle cell disease. It allows the Hct and HbS to be adjusted rapidly and simultaneously, without incurring the risks of increasing blood viscosity and blood volume. In theory, acute sickling episodes might be interrupted before tissue damage becomes irreversible.

Indications for exchange transfusion

The major role for exchange transfusion is in the management or prevention of life- or organ-threatening events. Unfortunately, randomised trials in such settings have rarely been possible; indications for exchange are therefore based largely on anecdotal reports.

Conditions for which relatively good evidence exists for the use of exchange transfusions include:

- **Acute chest syndrome (ACS)**
- **Acute or impending CVA**

Anecdotal success with exchange transfusion has been reported in the management of priapism failing to respond to 12-24 hours of conservative management with intravenous hydration, analgesia and supplemental oxygen.

Exchange transfusions may also be used prior to general anaesthesia in patients with a high baseline Hb (e.g. Hb SC disease) or a history of severe ACS or pulmonary disease.

Exchange transfusions can be used as an alternative to top-up transfusions for patients with sickle cell disease on regular transfusion programmes. This minimises iron overload, the main long-term complication of transfusion.
Before exchange transfusion

During working hours the decision to perform an exchange transfusion should be made by a consultant paediatric haematologist. Outside of working hours this decision should be made by the consultant on call for paediatric oncology/haematology in conjunction with the consultant on-call for adult non-malignant haematology if appropriate.

Confirm that blood has been sent for a full blood count, cross match, HbS%, coagulation screen, urea and electrolytes, creatinine, magnesium, calcium, phosphate, and liver function tests.

Calculate the amount of blood to be given, according to the formula below.

Inform the blood bank that you plan a red cell exchange in the next few hours and give them the patient’s name, date of birth, hospital number and location.

Send a request form to blood bank requesting the amount of blood required. State that the blood is for exchange transfusion on the form.

Red cells used for exchange transfusion should meet the specifications stated in the BCSH Transfusion guidelines for neonates and older children.

Discuss with the anaesthetist on call to organise insertion of a double lumen central venous line. If the child is old enough to tolerate the procedure without a general anaesthetic or is too unwell for a general anaesthetic, discuss with the paediatric intensive care medical staff to see if they are able to insert a central line.

Arrange for the child to be admitted to high dependency or intensive care for the exchange transfusion.
Methods

Manual

A single volume exchange will replace ~ 65% of the patient’s cells; a double volume exchange will replace ~85%. The target HbS level of below 30% may require repeated exchanges.

Practical Points

- Good venous access is essential:
  - Ideally a central venous line would be used for venesection with a connection via a 3 way tap to a red cell waste bag and a separate large bore cannula would be used for continuous infusion of replacement fluid.
  - Two cannulae will allow venesection from one and transfusion through the other.
  - An arterial line may be used for blood removal and a peripheral or central venous line for transfusion.
  - If it is only possible to get a single venous line, use a three-way tap to remove an aliquot of blood and then replace with the same volume (transfuse and venesect in aliquots of 10-30ml).

- Aim to exchange **0.5 – 1 blood volume in each exchange**

- As whole blood is being removed, and packed cells, with a higher Hct are being returned, if aiming for a final haematocrit equal to the starting haematocrit use the following calculation for a single volume exchange:
  - Total volume 80ml/kg
  - Volume of packed RBCs to be ordered = 50ml/kg
  - Volume of 0.9% saline = 30ml/kg

- If Hct < 0.2 begin by transfusing 10% of blood volume to avoid volume depletion.

- Venesect 5-10% of the patient’s blood volume using a 50ml syringe and replace with 0.9% saline – the same volume at the same time and rate to maintain isovolaemia (i.e. rate of removal equals rate of return)

- Subsequently, continue to remove aliquots of patient blood while maintaining isovolaemia with replacement fluid.

- For replacement fluid alternate a unit of RBCs with an appropriate volume of 0.9% saline to maintain the above ratio of RBCs to saline (5ml:3ml)

Close observation and monitoring of vital signs is essential during this procedure as fluctuations in blood volume can occur.

Following the procedure check FBC (including Hct) and HbS%, coagulation screen, urea and electrolytes, creatinine, calcium, phosphate and LFTs. If Hct > 0.35 additional saline may be required. The HbS level will guide the necessity for further exchange.
Automated

Modern cell separators e.g. COBE Spectra can perform automated red cell exchange.

This method, if available, is preferred to manual exchange – it is quicker and allows greater control of circulating volume.

Currently, automated red cell exchange is not available at Nottingham Children’s Hospital but patients over 40kg may be considered for exchange at City Hospital following discussion with the Consultant Haematologist.

Note: It is important that other aspects of the management of patients with sickle cell disease are continued when exchange transfusion is performed – i.e. re-hydration, adequate pain control, antibiotics, etc.

References


Sickle Cell Disease, Standards and Guidelines for Clinical Care in the UK (2nd Edition 2010)

Birmingham Children’s Hospital Haematology / Oncology Departmental Handbook (2006)

Guidelines for Transfusions in Children and Adolescents with Sickle Cell Disease, University Hospitals of Leicester Children’s Services (2010)