LRI Children's Hospital

Automated red cell exchange for paediatric sickle cell patients

Staff relevant to:	Paediatric haematology teams, paediatric medicine, paediatric intensive care
Team approval date:	April 2023
Version:	1
Revision due:	October 2024
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Trust Ref:	C19/2023

1. Introduction and Who Guideline applies to

This document aims to offer clinical guidance regarding automated red cell exchange transfusion in paediatric patients with sickle cell disease.

The policy applies to all patients with sickle cell disease who are starting/on a chronic exchange transfusion program and those who need emergency management of acute complications related to sickle cell disease.

Cases need to be assessed individually and management tailored appropriately. If in doubt, please seek the opinion of the paediatric haemoglobinopathy team.

Guidelines are based on:

- Sickle Cell Disease in Childhood: Standards and Recommendations for Clinical Care 3rd Edition 2019
- BSH Guidelines on red cell transfusion in sickle cell disease Part I: Principles and laboratory aspects & Part II: Indications for transfusion 2017

2. Sickle cell disease and automated red cell transfusion

2.1 General principles

The purpose of red cell transfusion in patients with sickle cell disease is to:

- Improve the oxygen carrying capacity by correcting anaemia.
- Prevent or reverse complications of sickle cell disease due to vaso-occlusion or haemolysis.

With exchange transfusion the aim is to remove the patient's sickle cells and replace with donor sickle-negative red cells so that the Hb S% is rapidly reduced to less than 30%, whilst maintaining a steady state blood volume throughout the procedure.

This is the optimal emergency management in acute sickle cell complications such as:

- Acute stroke
- Acute chest syndrome
- Fulminant hepatic failure
- Splenic or hepatic sequestration
- Fulminant priapism not responding to other therapies

Regular long-term transfusion consists of repeated red cell transfusions to keep the Hb S less than 30% over time. It is indicated for:

- Primary and secondary stroke prevention.
- Recurrent acute chest syndrome or painful episodes not prevented by hydroxycarbamide.
- Progressive organ failure.

2.2 Before starting transfusions

Transfusion can be a life-saving therapy for acute complications and reduce the risk of chronic progressive organ damage from ischaemic stroke but any decision to use transfusion therapy has to be weighed up against the potential risks and these should be discussed with the patient/family and documented in the notes. Patients with sickle cell disease receiving red cell transfusion are at particular risk of:

- Alloimmunisation resulting in red cell antibody formation. This can lead to a higher risk of delayed haemolytic reaction and make it more challenging to get appropriately cross-matched red cells.
- Hyperhaemolysis where there is bystander red cell destruction in the absence of antibody development. Management is individualised but should include avoiding further transfusion if possible. In severe cases early use of intravenous immunoglobulin and methylprednisolone is recommended as well as supportive care with recombinant erythropoietin, intravenous iron and B12/folate supplementation. With certain clinical criteria, ecluzimab is

commissioned for use in hyperhaemolysis but its use would need to be decided by a Consultant Haematologist.
- Increasing blood viscosity with an increase in overall haemoglobin in addition to Hb S containing cells.
- Secondary iron overload from chronic transfusion.

Before starting transfusions:

- Always discuss and document the indication for starting a red cell transfusion program.
- Potential complications of transfusion should also be discussed (use UHL blood transfusion consent sticker).
- All new patients should have a transfusion history taken and extended red cell phenotyping performed in addition to red cell grouping and antibody screen.
- Blood bank must be informed that the patient has sickle cell disease and requires sickle negative blood.
- Ensure patients are immunised against hepatitis B.
- Check hepatitis B, hepatitis C and HIV serology
- Written consent before starting red cell exchange programme then verbal consent at each transfusion.
- A decision made by the paediatric haemoglobinopathy team regarding desired Hb S%, haematocrit, frequency of transfusions and run time.
- Patients with haemoglobin of 60g/L or below will require a top up transfusion pre-exchange transfusion.

3. Managing the transfusion appointments

- Most patients will attend ward 27 day care at a pre-arranged time for pre transfusion sampling (FBC, G+S, U+Es, LFTs, ferritin, calcium, bone profile, magnesium and Hb S% for patients with sickle cell disease).

4. Automated red cell exchange process

4.1 Pre-Transfusion assessment:

- Check all above blood tests completed within the last 48 hours
- Baseline observations stable
- Height and weight
- Check appropriate venous access
- Check general health – if any general concerns discuss with appropriate consultant prior to starting procedure

Set up Terumo Spectra Optia for red cell exchange procedure and further discussion with consultant if depletion or depletion/exchange procedure is required.

4.2 Process during procedure:

- Monitor patient observations 5 minutes after the start of the exchange transfusion then after 5 minutes of commencing each unit of blood and more frequently if patient becomes unstable
- Document run times and machine values alongside observations with each unit of blood

4.3 Potential complications

- Managing citrate toxicity

a) Pause the system
b) Notify medical team of patient's condition
c) Options of treatment include slowing down inlet flow rate, reducing AC infusion rate. If symptoms still not alleviated, pause procedure, check calcium level and consider calcium replacement:

Hypocalcaemia guideline: **Calcium Disorders UHL Childrens Hospital Guideline**

- Vasovagal episodes

a) Pause the procedure
b) Lower the head of the patient and raise their feet
c) Restart once patient blood pressure is stable and consider reducing inlet flow rate to slow down procedure
d) If hypotensive, administer intravenous fluid bolus of 0.9% sodium chloride 10ml/kg and discuss with medical team to avoid hypervolemia
e) Consider monitoring the patient's observations more frequently

- Blood transfusion reaction

a) Manage transfusion related reactions as per UHL blood transfusion policy: **Blood Transfusion UHL Policy**

4.4 Post procedure

- End procedure without rinse back as this is not recommended in paediatric red cell exchange due to this procedure giving sickle cells back to the patient
- Record final run values on procedure flow chart
- Monitor patient's observations 15 minutes and 30 minutes after end of procedure.
- Recheck FBC and HbS %
- If patients well and observations stable after thirty minutes then flush port and lock with Heparin 100units/ml in total 4mls and remove access lines
- Ensure patient has follow up appointment for next blood tests and exchange transfusion

5. Monitoring for patients on chronic red cell transfusion

- All patients should have the following regular assessments at each transfusion visit and these will be documented in their electronic transfusion spreadsheet:
    - Weight
    - Pre-transfusion haemoglobin/haematocrit
    - Renal function
    - Liver function
    - Ferritin
    - Volume red cells transfused
- Iron chelation therapy will be reviewed every 3 months
- Patients with secondary iron overload/on chelation therapy will undergo regular ferriscan liver and cardiac T2* MRI to assess for iron loading in the heart and liver
- Patients on iron chelation therapy will undergo annual audiology and ophthalmology surveillance

- Viral serology for hepatitis B (Hep B surface Ag), hepatitis C (Hep C core antibody) and HIV (HIV I & II antibody) will be checked every 12 months as well as anti-Hepatitis B surface antibody
- Other relevant investigations related to endocrine, bone, cardiac and liver systems will be reviewed and requested at the time of annual review

6. Education and Training

Regular teaching provided on ward 27 medical and nursing days and within paediatric haemoglobinopathy team.

Only trained band 6 and 7 nurses signed off by a Terumo representative and who have completed the appropriate competencies required, will carry out red cell exchange procedures.

7. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Transfusion spreadsheets	3 monthly clinical review of patient and spreadsheets and annual review	Dr Kaljit Bhuller	3 monthly	Seen by haemoglobinopathy team

8. Supporting References

1) Sickle cell disease in childhood – standards and recommendations for clinical care 3rd edition November 2019

2) BSH Guidelines on red cell transfusion in sickle cell disease Part I: Principles and laboratory aspects & Part II: Indications for transfusion 2017

3) Calcium Disorders UHL Childrens Hospital Guideline 2020

4) UHL blood transfusion – policy and procedures for the prescribing, collection, storage and administration of blood and blood components 2021

9. Key Words

Sickle Cell, Anaemia, Exchange Transfusion, Haemoglobinopathy

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The Trust recognises the diversity of the local community it serves. Our aim therefore is to provide a safe environment free from discrimination and treat all individuals fairly with dignity and appropriately according to their needs.
As part of its development, this policy and its impact on equality have been reviewed and no detriment was identified.

CONTACT AND REVIEW DETAILS			
Guideline Lead (Name and Title) Kaljit Bhuller, Consultant in Paediatric & TYA Haematologist			**Executive Lead** Chief Nurse
Details of Changes made during review:			
Date	**Issue Number**	**Reviewed By**	**Description Of Changes (If Any)**
April 2023	**1**		**New document**