Exchange Transfusion procedure in children with Sickle Cell Disease (SCD)

Introduction

This document covers exchange transfusion procedure in children with Sickle Cell Disease.

Whenever possible, exchange transfusions will be undertaken by automated erythrocytapheresis by suitably trained nursing staff. This allows for safer, euvolemic and full-volume exchanges in a much quicker timescale compared to manual exchanges. Manual exchanges, however, may need to be undertaken out of hours, when trained staff is not available for automated exchanges or if suitable IV access is not secured.

Scope

This clinical guidance is for hospitals in the Thames Valley Haemoglobinopathy Network.

Procedure for automated emergency red cell exchange

Contact numbers:

- Oxford Medical Office 01865 387710 (daytime to find out which consultant is covering apheresis in Oxford)
- Daytime Transfusion Registrar OUH bleep 6888
- 24-hour Oxford Apheresis Unit and on call nurse 01865 387938
- 24-hour Regional Transfusion Medicine Consultant 01179594666 (use for out of hours apheresis requests and transfusion advice)

Phone to discuss all referrals with a doctor.

You will need to provide current history and reason for requesting exchange, transfusion history, HbS level and date, current height, weight, Hct in order for replacement blood volume to be calculated by TAS.

For information and referral forms: http://nssg.oxford-haematology.org.uk/
Select 'NHSBT' - this shows information documents and patient leaflets. Choose drop down menu and select 'referral to apheresis '. Use 'therapeutic apheresis' to request red cell exchange - all other procedure forms are here too in case you need them. Always use the website and you will have the current version as required by regulators.

Samples to send to Transfusion lab and process

- Send an urgent X match (full EDTA to the transfusion lab)
- All sickle cell patients known at CHOX may have had a red cell phenotype done by the lab if they have had a G and S.
- Those at CHOX who never had a group and save: the lab will do a red cell phenotype and antibody screen (takes 15 minutes)
- If from outside CHOX, the lab can either do this in house or they will contact the local lab for the phenotype/antibody screen.
- For full red cell exchange, and automated, for adults order 15 units, or children 8- 10 (lab will ask apheresis team how many units to order)
- Blood will be phenotype matched, HBS negative, less than 7 days old if possible
- Blood bank will take one hour in Oxford or 2-3 hours from Bristol.

Manual Red Cell Exchange
The following applies to manual exchanges only.

Exchange transfusion is undertaken to rapidly reduce the percentage of sickle cells in the circulation when a patient develops a life-threatening complication of the disease. It is not to be undertaken lightly, as the possible complications are considerable. However, patients with the following problems justify the risks:

- Severe chest syndrome (e.g. low or falling PaO_2) or Girdle syndrome
- A new CVA
- Multi-organ failure, e.g. associated with systemic fat embolism
- Fulminant priapism (> 4 hours) unresponsive to pharmacological therapy (as impotence can be the long-term outcome)

Exchange transfusions should only be undertaken by experienced medical and nursing staff and in an emergency situation and should not be initiated without discussion with the responsible consultant. The red cells used should be specially grouped (ABO compatible, Rh negative (rr) or RO as appropriate, Kell compatible), and HbS negative. Ensure the Blood Transfusion Laboratory is aware that the blood required is for exchange transfusion.

Aim of the procedure

a) To reduce the HbS level to < 20% over 2 - 3 days unless acutely ill, when more rapid exchange may be appropriate
b) To keep Hb $\leq$ 100 g/l initially and by the end of the whole procedure (or at steady state level in those with higher baseline Hb, e.g. HbSC patients 110-120 g/l).
c) To maintain a steady state blood volume and Hb throughout the exchange

- Use SAG-M blood, which is the freshest available (to prolong its life in the patient).
- Do not use diuretics.
- Continue to administer IV fluids at the standard rate between transfusions.

If manual exchange is being used, critically ill patients may require them more frequent than daily for rapid reduction of sickle percentages. Where possible, leave a 4 - 8 hour break between manual exchanges. For patients undergoing exchanges, particular attention should be paid to PaO_2, CVP, acid-base balance, U&E, Ca^{++}, citrate load, core temperature and clotting.

Preliminary investigations

- FBC
- HbS level (or S+C level if HbSC disease), if not previously available. Hb S levels do not fluctuate significantly in patients who are not transfused and so pre-transfusion Hb S levels are not necessary in an emergency in un-transfused patients known to the paediatric haematology service.
- Extended RBC phenotype (if not already known), x-match.
- U & Es, Ca^{++}
- Venous blood gases - in those with symptoms suggestive of acute chest or girdle syndrome,
- Serology for Hepatitis B & C, if not done recently.

Volumes required

The number and volume of exchange transfusions performed in a child with sickle cell disease will depend on the severity of the clinical problem and the haemodynamic stability of the child

A 'total' exchange is 1.5 - 2 times the child's blood volume and this is usually performed in 3 or 4 exchange procedures

Volume (ml) of blood removed *for each procedure* should be:

30 x weight (in kg) = volume in ml

Preparation of the patient

Patients must be well hydrated prior to starting an exchange transfusion. Adequate explanation must be given to the child and parents as to the indication for exchange and its potential risks and benefits and this should be documented in the case-notes.

Venous access

Two ports of venous access are required; one for venesection, the other for administering blood and crystalloid; in certain circumstances, an arterial line may be used for venesection.

Procedure

- The aim is that this should be an isovolaemic procedure with monitoring of blood pressure, heart rate and oxygen saturations every 15 minutes, and 1 hourly temperature monitoring. Exchanges are done in 'aliquots' of approximately 1/10 of the total to be exchanged (never greater than 5% of the blood volume). Note that the haematocrit of transfused packed cells (approximately 0.5-0.7) is higher than that of the venesected blood.

- It is important to ensure that the child is well hydrated between successive exchanges and that the haemoglobin is regularly monitored and kept at or below 100 g/l at all times.

- For the first procedure, start by venesecting 30% of the above volume (ie approx 10 ml/kg) in aliquots of 10 - 50 ml using a large syringe. Normal saline should be concurrently infused at the same rate to maintain isovolaemia. If the child has haemoglobin of less than 60 g/l replacement with blood rather than normal saline will be needed. *It is essential to ensure that blood is ready and easily available from Blood Bank before commencing.*

- The venesected blood can be discarded into a venesection bag via a 3-way tap. Venesection bags are kept on PHD unit.

- Continue venesecting the remaining 70% of the volume (approx.. 20 ml/kg) replacing with blood at a rate of 10 ml per kg per hour (red cells in SAG-M) instead of normal saline. This process should take about 120 min, depending on the rate of flow of blood and the clinical condition of the patient.
- Where possible leave at least 4-6 hours between each exchange procedure. In critically ill patients exchanges may need to be continuous.

- Check FBC, HbS %, U&E including calcium, and clotting, at the end of each procedure. Ensure that the Hb $\leq$100 g/l to reduce the risk of hyperviscosity.

- Continue with 3 to 4 exchange procedures until the Hb S <20%.

- NB For the second and subsequent exchange procedures, it may be necessary to use a ratio of 50% saline: 50% blood (rather than 30:70) in order to prevent the Hb from rising too high. This should be discussed with the paediatric haematology SpR or consultant when planning the procedure.

- A top-up transfusion can be given at the end of the final exchange procedure to give a final Hb of 100 g/l.

Possible immediate complications

- Anaphylactoid reactions due to HLA antibodies. Treat with antihistamine and/or corticosteroid
- Metabolic disturbances are rare, occurring usually in small children, or in association with visceral sequestration requiring continuous exchange.
- Convulsions are very rare. They are usually a sign of cerebral sludging, often in patients with previous CNS problems. Check that Hb has not risen too high (> 110 g/l). Give anti-epileptics; ensure there is a large fluid intake; give oxygen.
- Hypertension is occasionally seen in patients with circulatory overload. If diastolic BP increases by > 20 mmHg, slow down exchange, check Hb not >110 g/l or hct not > 0.4. If diastolic BP is > 100 mmHg stop the exchange, venesect, and consider antihypertensives.

Exchange transfusion (example)

A total exchange is 1.5 - 2 times the child's blood volume. This is normally performed in 3 - 4 exchange procedures. Each exchange procedure should be calculated at

30 ml/kg A child of 25 kg x 30= 750 mls

Childs Hb 70 g/l thus start with saline
10 ml/kg = 250 ml saline in and 250 ml
blood out
Then 20 ml per kg of blood = 500 ml blood in and 500 ml out

N.B. If starting Hb below 60 g/l all blood should be used = 750 ml blood in and 750 ml blood out. Record in 10 - 15minute cycles.

For example

Time	Saline in	Blood in	Blood out	Balance
10.00-10.10	31		31	31 in 31 out
10.10-10.20	31		31	62 in 62 out
10.20-10.30	31		31	93 in 93 out
10.30-10.40	31		31	123 in 123 out
10.40-10.50	31		31	154 in 154 out
10.50-11.00	31		31	185 in 185 out

This example is typical of that required for a clinically unstable patient; slighter higher rates may be used in patients who are cardiovascularly and neurologically stable (see main text for further information on volumes and overall timing).

Exchange Transfusion Form

Time	Saline in	Blood in	Blood out	Balance

This is a controlled document and therefore must not be changed

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Review

Name	Revision	Date	Version	Review date
Amrana Qureshi	Routine Review	July 2021	1.3	July 2023
Manuela Sultanova	Inclusion of scope, introduction, review and authors. Updating the logo and formatting the document	Feb 2022	1.4	July 2023
Amrana Qureshi	Full Review	October 2023	2.0	October 2023