Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric guideline

Exchange Transfusion

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 and when a simple 'top up' transfusion is deemed not appropriate in the particular clinical scenario. The decision to proceed with an exchange transfusion should be taken following discussion with the Paediatric Haematology Consultant on call as the procedure is not without possible complications. Indications for exchange transfusion in patients with sickle cell disease include:

- Severe acute chest syndrome – see ACS protocol for indications
- Girdle syndrome
- Acute stroke (or as part of primary/secondary stroke prevention)
- Multi-organ failure syndromes
- Electively, in preparation for planned surgical procedures in selected cases

Aims

i) To reduce the % HbS to < 20% over 2-3 days unless acutely ill, when more rapid exchange may be appropriate

ii) To keep Hb < 100g/l initially (or at steady state level in those with higher baseline Hb, e.g. HbSC patients) and about 100-110g/l (PCV 0.36) by the end of the whole procedure.

iii) To maintain a steady state blood volume throughout the procedures

Preliminary Investigations

- FBC
- % HbS (or S+C if HbSC disease)
- Extended RBC phenotype (if not already known), cross-match and antibody screen (ensure that the blood transfusion laboratory is aware that the blood requested is for a child with a haemogobinopathy AND for exchange transfusion so unit specifications can be met)
- urea and electrolytes, creatinine, calcium
- Capillary or arterial blood gases - in those with symptoms suggestive of chest or girdle syndrome
- Baseline Serology for Hepatitis B and C and HIV, if not done recently

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Procedure

- Use SAG-M blood, which is the freshest available (to prolong its life in the patient)
- Red cells should be phenotype compatible i.e. ABO, Kell and Rh compatible (rr or Ro as appropriate)
- Do not use diuretics
- Continue to administer IV fluids at the standard rate between transfusions

Critically ill patients may require exchanges to be more frequent than daily. Where possible, leave a 4 - 8 hour break between exchanges. In the very sick patient, the procedure is a continuous process. In these patients, particular attention should be paid to PaO_2, CVP, acid-base balance, Ca^{++}, citrate load, core temperature and clotting.

Automated Exchange Transfusion

A national SNBTS SOP for automated red cell exchange exists and is available through SNBTS.

- Information required:
- Patient height and weight
- Pre-exchange Hb, Hct and HbS%
- Transfusion fluid Hct (or default 60%)
- Target values post red cell exchange
- Hb 100g/l
- Hct 0.35
- HbS 15%
- FCR – fraction of red cells remaining – can be calculated on apheresis instrument from pre and target post HbS %

Adequate venous access is required. If it is anticipated that adequate access may not be obtained via peripheral access then insertion of a large bore double lumen catheter with staggered ends should be arranged.

Manual Exchange Transfusion

Volumes Required

The initial aim is to exchange 1.5 - 2 times the child's blood volume, divided over 2-3 procedures. 28ml/kg is the approximate red cell mass from infancy to teenage years

Volume (ml) of SAG-M blood *for each exchange* should be:

28 x weight (in kg) = volume in mls

Venous Access

Two ports of venous access are required; one for venesection, the other for administering blood and crystalloid;

Procedure

The aim is that this should be an isovolaemic procedure with frequent, 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.

- If Hb >60g/L start by giving a bolus of 10ml/kg of 0.9%saline. If Hb < 60g/l, start by transfusing 5ml/Kg of red cells then proceed to venesection.
- Venesect an aliquot (i.e. 10% of total volume to be exchanged) over 10- 15 minutes
- Transfuse an equal first aliquot at a rate of 5-7.5ml/kg/hr
- Venesect second aliquot
- Transfuse the second aliquot
- Venesect third aliquot, and so on
- Finish the procedure by giving the last aliquot of blood as a top-up transfusion ie venesect the 9th aliquot then transfuse 9th and 10th aliquots

At end of procedure check FBC, HbS %, urea and electrolytes including calcium. If HbS not <20%, then consider continuing with further exchanges, to give a final Hb of 110g/l and Hb S ideally between 10% and 20%.

Ensure the child is well hydrated between successive exchanges as the haematocrit of transfused packed cells is higher than that of the venesected blood. Keep PCV<0.4. In larger volume exchanges consider giving a break between 2nd or 3rd unit and giving dextrose/saline to rehydrate.

Possible Immediate Complications

Transfusion reactions - ABO incompatibility, febrile non-haemolytic reactions, TRALI etc

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 the PCV has not risen too high (>0.4). 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 > 20mmHg, slow down exchange, check PCV not >0.4. If diastolic BP is >100mmHg stop the exchange, venesect, and consider antihypertensives.

NOTE

This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.