Introduction
This document aims to offer guidance in association with transfusion practices in relation to Sickle Cell patients.
The policy applies to all staff responsible for patients with Sickle Cell and Thalassaemia.

General Principles

- Blood transfusion should only be used for specific indications.
- Blood transfusion should be used as sparingly as possible.
- Avoid hyperviscosity by ensuring final haematocrit <0.35.
- Give sickle negative, CMV negative, phenotypically matched blood.
- Transfusion will NOT reduce the severity or duration of uncomplicated painful crises.

Investigations

For all transfusions in patients with sickle cell disease it is important to record the following information before and after the procedure:

Hb
WCC
Platelets
Hct
%Hbs
U&E
Ca
LFTs

Top Up Transfusion

Indications

When a patient with SCD drops their Hb sufficient to cause, or risk, clinical compromise such as cardiac failure and hypovolaemic shock.

It is rarely necessary to transfuse if the Hb is > 5g/dl, or <2g below steady state unless there is reticulocytopenia associated with the falling Hb or clinical evidence that the fall in Hb will continue.

Top up transfusion may also be indicated in patients with moderately severe crises, where exchange transfusion is not felt necessary.
These circumstances may arise with:

- Splenic sequestration.
- Hepatic sequestration.
- Acute chest syndrome.
- Mesenteric syndrome.
- Aplastic crises (Parvovirus B19.)
- Blood loss.
- Haemolytic crises.

Other possible indications:

- Pre-operative.
- Pre-flying.

**Aims**

To improve oxygen carrying capacity.
To dilute sickle cells and improve blood viscosity and flow.

Raise Hb to patient's steady state level.
Do not raise Hn > 10g/dl (or Hct>0.35) so as to avoid increasing blood viscosity.

**Practical Points**

Volume of blood to be transfused (mls) = (desired Hb – current Hb) x weight (kg) x K.

K is a constant depending on the haematocrit of the blood to be transfused:
- 3 – packed red cells
- 4 – plasma reduced blood (SAG-M)

In severe anaemia do not transfuse >5ml/kg at one session or >2ml/kg/4 hours.

**Blood Products**

Usually packed red cells are used.
These should be ABO and Rh compatible, CMV negative and sickle negative.

**Exchange Transfusion**

**Indications**

Exchange transfusion is generally reserved for the treatment of life- or organ- threatening and other major complications of SCD.

These include:

- Severe acute chest syndrome.
- Mesenteric syndrome.
- CVA.
- Splenic sequestration.
- Hepatic sequestration.
- Fulminant hepatic failure.
- Priapism unresponsive to therapy.
Other possible indications:

- Pre-operative.
- Pre-flying.

Aims

1. To improve oxygen carrying capacity.
2. To remove sickle cells, dilute those remaining and improve blood viscosity and flow.
3. To avoid a rise in haematocrit and blood viscosity (final Hct<0.35).
4. To reduce final HbS to <30%.
5. To keep final Hb <12.5g/dl.
6. To avoid changes in circulating blood volume.

Methods

a) Manual

Volume of blood to exchanged = 1.5 – 2 x total blood volume (80ml/kg)

(A single volume exchange will replace ~65% of the patient’s cells; a double volume exchange will replace ~85%).

This method is time-consuming and it may not be possible to achieve the desired reduction in HbS% in one procedure. It is usual that 3-4 exchanges will be necessary lasting 2-4 hours each.

Practical Points

Aim to exchange 0.5 – 1 blood volume in each exchange.

Venesect 5-10% of the patients blood volume and replace with 0.9% saline – the same volume in the same time and rate to maintain isovolaemia. Continue to exchange with donor blood, venesecting and transfusing at the same rate.

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

Good venous access is essential:

- Two cannulae will allow venesection from one and transfusion through the other.
- In some patients a central venous line may be required – this can be used, with a three-way tap, for the exchange transfusion.
- 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.)

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, U&Es, Cr, Ca, PO4, albumin, and LFTs. This will guide the necessity to perform a second exchange.

b) 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. Access to automated red cell exchange will depend on the time of day and the availability of trained staff to operate the cell separator machine. Please discuss with Consultant Haematologist.

Practical Points

Good venous access is essential in order to maintain adequate flow rates, which are necessary to the machine running. Temporary central venous lines may be necessary in some patients.

Volume of blood to be exchanged is calculated by the machine but should equate to 1-1.5 x total blood volume (80ml/kg).

Information required pre-exchange.

Patient details.
  Sex.
  Height.
  Weight.
  Pre-Hct.

Hct of transfused blood.
  0.65 for packed red cells (0.55-0.75).
  0.6 for SAG-M blood (0.5-0.7).

Desired post Hct.
  0.35 (to avoid hyperviscosity).

Desired % FCR (fraction of patient cells remaining)
  20%.

Fluid balance.
  0-10%.

During automated red cell exchange patient may experience symptoms related to hypocalcaemia (peri-oral paraesthesia, tetany etc). These can generally be prevented by administration of oral calcium supplements during the procedure. If they occur they can be relieved by administration of intravenous calcium gluconate (0.5ml/kg 10% calcium gluconate).

There may be loss of platelets during these procedures. It is therefore important to check FBC following the procedure.

Blood needs to be warmed before transfusion.
Blood Products

Usually packed red cells or SAG-M are used. These should be ABO and Rh compatible, CMV negative and sickle negative. Do not use diuretics with the transfusion – risk of increasing blood viscosity.

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

Hypertransfusion

Definition: Repeated transfusion to keep Hbs <30%.

Patients will be considered for hypertransfusion regimens on an individual basis.

To hypertransfuse:

Transfuse at 3 – 4 week intervals to suppress erythropoiesis and keep HbS <30%.
Aim for Hb 11-13 and no >14g/dl after transfusion.
Patients vary in the frequency and amount of blood required to suppress HbS production. In children with SC disease it is usually necessary to start with an exchange. Transfusion and in other children in it may be necessary to exchange at times.

Pre-operative transfusion

The decision to transfuse before surgery needs to be considered on an individual basis. Many procedures may be performed without the need for transfusion. If transfusion is considered necessary the usual aim is to reduce the Hbs level to <30%. This can be done by top up transfusion or exchange.

Please discuss all cases with the Consultant Haematologist in charge of the patient.

Specific complications of blood transfusion in sickle cell disease

1. Hyperviscosity

This may result in fitting, poor cerebral function and a worsening clinical condition.
Check the Hct is <0.35.

2. Metabolic disturbance

These include hyperkalaemia, hypocalcaemia, citrate toxicity, Hypernatraemia, hypo – or hyperglycaemia and late onset alkalosis. Be aware of these complications and check biochemical profile after the procedure and correct as necessary.

Legal Liability Guideline Statement

Guidelines issued and approved by the Trust are considered to represent best practice. Staff may only exceptionally depart from any relevant Trust guidelines providing always that such departure is confined to the specific needs of individual circumstances. In healthcare delivery such departure shall only be undertaken where, in the judgement of the responsible healthcare professional’ it is fully appropriate and justifiable - such decision to be fully recorded in the patient’s notes.