Blood Transfusion – Children requiring regular treatment - Full Paediatric Clinical Guideline – Joint Derby and Burton

Reference no.: CH CLIN G111

Purpose

To improve and standardise care of children who require regular blood transfusion for disorders such as Sickle Cell Disease or Thalassemia.

Aim & Scope

This guideline covers the management of children having regular blood transfusions on the day case ward and paediatric wards.

Indications for regular transfusion

Children having regular blood transfusions include those with:

- β Thalassemia major
- β Thalassemia intermedia
- Children with rare inherited causes of anaemia including unstable hemoglobin's & Diamond Blackfan Anaemia
- Some children with Sickle Cell Disease

The aim of regular transfusion in sickle cell disease is to maintain the level of HbS less than 30%. Regular transfusions may be used in the following situations:

- Secondary prevention of recurrent strokes after an initial CVA
- Primary prevention of stroke in patients shown to be at risk by having elevated transcranial Doppler velocity
- In young children with sequestration crises in an attempt to delay or avoid splenectomy
- In patients with recurrent acute chest syndrome, chronic pulmonary disease and symptomatic congestive heart failure
- Selected patients with severe, recurrent, debilitating symptoms such as painful crises, leg ulcers, and priapism may benefit from short periods of regular transfusion.

Investigations and vaccinations prior to first transfusion

- Any children not vaccinated against hepatitis A and B should be offered immunisation.
- Antibodies to hepatitis B, hepatitis C, and HIV should ideally be determined beforehand.
- Ensure that an extended red cell phenotype has been performed.
- Check baseline LFTs and ferritin

Designated area for transfusion

Transfusions will usually be given on Children's Daycare area.

If a child on a regular transfusion programme is an inpatient, when their transfusion is due, the transfusion may be given on one of the paediatric wards.

Pre-transfusion assessment / bloods

Blood samples need to be taken 1-3 days prior to transfusions

Blood should be taken for (*check Lorenzo and attach samples*):

- Antibody screen and cross match (written form)
- Full blood count (needs separate sample to the cross match sample)
- In sickle cell disease- HbS% monthly until < 30% then 3 monthly (order as Haemoglobin electrophoresis)
- LFTs and Ferritin monthly
- Children taking Deferasirox (Exjade) need LFT, U&E and Ferritin monthly for monitoring.
- Further information on children on iron chelation can be found within the paediatric guideline: Iron Chelation Therapy when Receiving Regular Blood Transfusions on the Trust Intranet

Transfusion

Prior to each transfusion a full set of observations (temperature, heart rate, blood pressure, respiratory rate and oxygen saturation) should be performed.

The nurse who will be setting up the transfusion will follow all procedures specified in the Trust Transfusion Policy *(See paragraph below).* They should check with the family/child that the child is not unwell.

If there are abnormal observations or there are concerns that the child is unwell the child should be reviewed by a doctor who will decide if they are fit for the transfusion.

Dr Weights will have already made the decision to start regular transfusions and will go through the process before transfusions start. A specific transfusion plan will be available for each patient. **It is imperative to check this plan each time for any changes or specific requirements for each patient.**

If the patient has a 'Portacath' this will be accessed by one of the day care nurses competent in this technique. If they do not have a Portacath then they will need to be cannulated for the transfusion

No more than 2 attempts at cannulation should be made by 1 individual.

Rarely, if cannulation is difficult and the child is becoming distressed the decision should be made to abandon the transfusion and bring the child back another day.

The transfusion should aim to be started within 1 hour of the child's arrival to Day Care.

Requirements for blood products

- Packed red cells which are fully Rh and Kell compatible.
- If alloantibodies are identified, further transfusions should be negative for corresponding antigen.
- Red cells for transfusion to patients with sickle cell disease should be sickle-test negative
- Red cells should ideally be less than 7 days old

Transfusion Policy

The following policy is available via the trust intranet and provides guidance for all transfusions:

TRUST POLICY AND PROCEDURES FOR THE TRANSFUSION OF BLOOD AND BLOOD COMPONENTS (Reference CL-RM/2012010)

The volume of packed red cells to be transfused should be calculated according to the formula below:

Volume to be Transfused (mls)	=	Weight of the patient (kg)	X	Aimed for increment of Hb (g/L)	X	0.4

Example: Child weighing 35Kg with an Hb of 105 would require 35x35x0.4 = 490mls of packed red cells to increase Hb to a target of 140g/L

Do not give more than 20ml/kg per transfusion.

Blood should be given over a period of 3 to 4 hours. The maximum flow rate is 5mls/kg/hour

Blood should be transported from blood bank in a transport box with cool packs. The transfusion should be set up as soon as the blood arrives on the ward and given over 4 hours. Please ensure that the cool box is returned to blood bank in a timely manner.

Each child on regular transfusions will usually have their own individual transfusion plan in the folder on day case or in their notes. Each child will have their own target Hb - this is usually 140 g/L but may be 120 g/L therefore please check individual management plans to be sure of the target Hb.

For children with Thalassemia the target Hb should not exceed 140g/L.

When a transfusion program is initiated, Dr Weights will decide on the target Hb and will write an individual management plan for the patient.

The volume of blood transfused should be adjusted to:

- Maintain the pre-transfusion haemoglobin levels above 95-100g/L in patients with thalassaemia
- Maintain the HbS < 30% in patients with sickle cell disease.

Adjustments to the volume of blood to be transfused should only be made after discussion with the patient's consultant paediatrician (Dr Weights).

Risks associated with regular transfusions

The risks associated with regular transfusion include acute transfusion reactions, allo-immunisation to red cell antigens, transmission of viral infection, and, in the long term, iron overload. Acute transfusion reactions should be managed according to the Trust Transfusion Policy.

Frequency of transfusions and further appointments.

Transfusions should generally be given at an interval of 4 weeks unless specified by the Consultant Paediatrician.

Transfusions should be scheduled in advance and maintained at a fixed schedule. This enables patients and families to establish routines and will improve quality of life.

All families should receive forms for cross match and monitoring bloods prior to leaving from their transfusions. Appointments should be made for the next transfusion and blood tests. Cross matches are valid for 72 hours in patients on regular transfusions.

Transfusions for older children should be offered out of hours to minimise disruption to the child and their family. However, it is important to maintain regular review of these children by the specialist team.

References (including any links to NICE Guidance etc.)

- Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK (2008)
- Standard of Care Guidelines for Thalassaemia. Children's Hospital & Research Center Oakland (2008)
- Guidelines for Transfusions in Children and Adolescents with Thalassaemia Major. University Hospitals of Leicester NHS Trust Children's Services (2010)
- Guidelines for Initiation of Blood Transfusion in New Patients with Thalassaemia Major/ Intermedia. University Hospitals of Leicester NHS Trust Children's Services (2010)
- Guidelines for Transfusions in Children and Adolescents with Sickle Cell Disease. University Hospitals of Leicester NHS Trust Children's Services (2010)
- Sickle Cell Disease in Childhood, Standards and Guidelines for Clinical Care in the UK (2nd Edition 2010)
- Adams RJ, McKie VC, Hsu L, Files B, Vichinsky E, Pegelow C, Abboud M, Gallagher D, Kutlar A, Nicols FT, Bonds DR, Brambilla D. Prevention of a first stroke by transfusions in children with sickle cell anemia and abnormal results on transcranial Doppler ultrasonography. N Engl J Med. 1998;339(1):5-11.
- Guidelines for In-patients Management of Children with Sickle Cell Disease (The Hospital for Sick Children Toronto-Canada (2006))

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