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SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric Guideline - Chronic Transfusion in children with Sickle Cell disease

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.

Indications for chronic transfusion

- Primary prevention of stroke – in children age 2-16yrs with abnormal TCDs.
- Prevention of recurrent stroke.
- Secondary prevention of silent cerebral infarction (SCI) severe disease i.e. recurrent painful vaso-occlusive crises, recurrent acute chest syndrome or chronic organ damage in children in whom hydroxycarbamide has failed, is contraindicated or unacceptable.
- Transfusion for indications where evidence is limited should be based on a case-by-case assessment after full risk-benefit analysis.

Transfusion for cerebrovascular disease

- Regular red cell transfusion to maintain HbS level <30% is indicated for the primary prevention of stroke in children (2–16 years) with SS or S/Beta^0 thalassaemia with abnormal TCD velocities. (STOP study).
- The TWiTCH study demonstrated that after transfusion for 1 year, for children established on the maximum tolerated dose of hydroxycarbamide and without severe vasculopathy, transfusion be safely discontinued with appropriate monitoring.
- The SIT study (2014) demonstrated a benefit from regular transfusion in children age 5-15 yrs of age with HbSS or HbS/beta^o who had evidence of silent cerebral infarcts. Treatment options including transfusion should be discussed with families of children who are found to have silent cerebral infarcts.
- Long-term transfusion to maintain HbS <30% (after is recommended for the prevention of recurrent ischaemic stroke due to sickle cell disease in children and stem cell transplant should be considered.

Pre-Transfusion issues

- Ensure that hepatitis A and B vaccination has been carried out with an adequate response.
- Ensure that extended RBC antigen phenotyping has been performed.
- Blood group genotyping should be considered in children with SCD who develop alloantibodies or who start a long-term transfusion programme.
- The choice of transfusion method, i.e., simple (top up) or exchange, should be individualised, taking into account the indication for transfusion, the need to avoid hyperviscosity and minimise alloimmunisation, maintenance of iron balance, venous access issues and available resources.

Red cell requirements

- ABO compatible, extended Rh and Kell matched.
- Antigen negative for current and historical clinically significant red cell antibodies.
- Units should be < 7days old for exchange transfusion or < 10 days old for top-up transfusion (older is permissible if there are matching issues).
- HbS negative.

Transfusion volume: In order to avoid hyper-viscosity post transfusion haemoglobin levels should be individualised to take account of the pre-transfusion HbS level.

NB. Patients should not wait longer than 30 minutes to commence a pre-arranged transfusion.

Monitoring

- Regular height & weight (3 monthly).
- Record transfusion requirements – volume, frequency and target/actual pre transfusion Hb and HbS%.
- Monitor for evidence of iron overload (refer to Iron Overload and Chelation Therapy Guideline that is available on the SPAH website).
- Children being transfused for cerebrovascular indications will require follow up TCD +/- MRI/MRA monitoring.
- SCD patients on regular transfusions should be screened annually for hepatitis B, hepatitis C and HIV.

References

1. BSH Guidelines on red cell transfusion in sickle cell disease. Part I: principles and laboratory aspects.
2. BSH Guidelines on red cell transfusion in sickle cell disease Part II: indications for transfusion.
3. Sickle Cell Disease in Childhood: Standards and Recommendations for Clinical Care 3rd Edition November 2019.
4. Hydroxycarbamide versus chronic transfusion for maintenance of transcranial Doppler flow velocities in children with sickle cell anaemia – TCD With Transfusions Changing to Hydroxyurea (TWiTCH): a multicentre, open-label, phase 3, non-inferiority trial. Russell E Ware et al. The Lancet Vol 387 Feb 13, 2016 p 661-670
5. Controlled Trial of Transfusions for Silent Cerebral Infarcts in Sickle cell Anaemia. MR DeBaun et al N Engl J Med 371;8 August 21 2014 p699-710