NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Paediatric guideline

Chronic Transfusion in Thalassaemia

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined based on 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.

This guidance has been prepared by NHS National Services Scotland (NSS) National Networks. Accountable to Scottish Government, NSS works at the heart of the health service providing national strategic services to the rest of NHSScotland and other public sector organisations to help them deliver their services more efficiently and effectively. Working across professional and organisational boundaries, National Networks support the delivery of safe, effective healthcare that's designed around patients, carers and families.

Contents

Authors	S Chaudhury, Consultant Paediatric Haematologist, NHS Greater Glasgow & Clyde S Baird, Consultant Paediatric Haematologist, NHS Lothian
Stakeholders involved	Consultant Paediatric Haematologists, Consultant Paediatricians, Haematology Nurse Specialists, Consultant Paediatric Radiologist
Others consulted	J Laird, Consultant in Transfusion Medicine, NHS Greater Glasgow & Clyde N Priddee, Consultant Haematologist, SNBTS
Methodology used	Evidence-based literature review and engagement with clinical experts within the field of Paediatric Haematology and Transfusion.
Rationale	The network was established to support and develop haemoglobinopathy services throughout Scotland in improving standards of clinical care for patients. The network supports the delivery of evidence based, patient centred care through the development and implementation of clinical guidelines, care pathways and information resources utilising a once for Scotland approach. Document first created in 2014. To aid healthcare professionals in NHS Scotland in clinical decision making about the appropriate and effective care of patients with thalassaemia who require regular transfusion.
Scope	Haematology specialists and general paediatricians managing children with haematological conditions in NHS Scotland where patients are transfusion dependant.
Approval process	The guideline was approved by the SPAH Paediatric Guideline and Protocol subgroup on 25 April 2025. This subgroup has the authority to develop, review and endorse guidelines. This has been agreed by the SPAH Steering Group and noted in its Terms of Reference.

	See appendix for list of Paediatric Guideline and Protocol subgroup members

Decision to start regular transfusion

Transfusion should be started when:

- there is significant anaemia Hb <70g/l on 2 occasions >2 weeks apart

AND / OR

- significant symptoms of anaemia
- poor growth / failure to thrive
- complications from excessive intramedullary haematopoiesis such as pathological fractures and facial changes

Aims of transfusion

The aim of blood transfusion in thalassaemia is to deliver a safe and effective transfusion regimen whilst minimising the burden of transfusion therapy. An effective transfusion regimen will result in:

- good growth and development
- good energy levels
- sufficient suppression of intra and extramedullary haematopoiesis

Trough Hb levels should be maintained at 95-105 g/l
Consider a higher target trough of 110-120g/l if cardiac issues, extramedullary haemopoiesis or other medical conditions.
A period of hyper transfusion (trough >130 g/l) may be considered prior to SCT.

Pre transfusion issues

- extended RBC phenotyping or genotyping should be undertaken before starting regular transfusion and should include typing for: D, C, c, E, e, K, Fya, Fyb, JKa, JKb, M, N, S and s
- vaccinate against hepatitis B
- informed consent for long term transfusion should be recorded and reviewed on a regular basis

Red cell requirements

- ABO compatible
- fully matched for Rh (CcDEe), and K phenotype
- negative for current and previously detected clinically significant antibodies
- where possible freshest available

Transfusion

- pre-arranged transfusion should be started within 30 minutes of arrival
- a transfusion volume of 15-20ml/kg every 3-4 weeks should achieve the required aims with a usual post transfusion target of 130-150g/l
- transfusion volume - can be calculated using the formula
    - volume to transfuse (mL) = [desired Hb (g/L) – actual Hb (g/L)] x weight (kg) x 0.3-0.4
- do not start an additional unit for a small amount of blood as this is wasteful and increases donor exposure.
- rate of transfusion - Transfuse at a rate of 5ml/kg/h, maximum 150ml/h *(NHSBT 2020)*

Monitoring

- regular height & weight and assessment of spleen size (3 monthly)
- record transfusion requirements (volume and frequency), pre-transfusion Hb, venous access attempts and transfusion reactions at each transfusion
- review red cell antibodies, transfusion reactions, venous access and annual transfusion requirements at regular/annual review.
- calculate annual transfusion requirements ml/kg/year based on mid-year weight. If annual requirements are >200ml/kg/year, then splenectomy may be considered to reduce transfusion requirements.
- transfusion reactions should be investigated and managed according to the BCSH guideline on the investigation and management of acute transfusion reactions.
- monitoring for evidence of iron overload according to SPAH Paediatric Iron Overload and Chelation guideline

References

Standards of the Clinical Care of Children and Adults living with Thalassaemia in the United Kingdom. 4th Edition (2023)

Position paper on International Collaboration for Transfusion Medicine (ICTM) Guideline ‘Red blood cell specifications for patients with hemoglobinopathies: a systematic review and guideline’ Sara Trompeter, Edwin Massey, Susan Robinson, on behalf of the Transfusion Task Force of the British Society of Haematology Guidelines Committee Vol 189 (3), May 2020

Appendix - Guidelines and Protocols subgroup membership

Name:		Job Title:	Organisation:
Katrina	Adams	Non-Malignant Haematology Nurse Specialist	NHS GG&C
Ruth	Allen	Consultant Paediatric Radiologist	NHS GG&C
Susan	Baird	Consultant Paediatric Haematologist	NHS Lothian
Nadia	Catherwood	Paediatric Haematology Nurse Specialist	NHS GG&C
Shahzya	Chaudhury	Consultant Paediatric Haematologist	NHS GG&C
Sarah	Clarke	Non-Malignant Paediatric Haematology Consultant	NHS GG&C
Emma	Cockburn	Consultant Paediatrician	NHS Tayside
Cheryl	Hill	Non-Malignant Haematology Nurse Specialist	NHS GG&C
Kirsten	Husselbee	Consultant Paediatrician	NHS Tayside
Jennifer	Milne	Paediatric Haematology Nurse	NHS Tayside
Lucy	Paterson	Clinical Nurse Specialist - Benign Haematology	NHS Lothian
Fernando	Pinto	Consultant Paediatric Haematologist	NHS GG&C
Gordon	Taylor	Consultant Paediatric Haematologist	NHS Grampian
Rosina	Shujaat	Consultant Paediatric Haematologist	NHS Lothian