Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric Guideline - Inpatient Specialist Management of acutely unwell children with haemoglobinopathies

Background
Children with sickle cell disease and thalassaemia have complex care needs. There is a high mortality associated with some of the acute complications of sickle cell disease and Thalassaemia. These children can deteriorate rapidly and an awareness of potential complications, their appropriate management and prompt access to specialist care including facilities for red cell exchange and critical care is imperative when managing these cases.

Transfer pathway from referring hospitals to tertiary referral centre

- Patients in centres without PICU and/or automated red cell exchange facilities should be transferred to RHCYP Edinburgh or Glasgow RCH depending on availability of critical care beds and specialist expertise. Preference should be given to transferring patients to centres where they are already known to the haematology team.
- **In all cases the on-call paediatric haematology consultant at the tertiary referral centre will discuss the patient with specialist teams, critical care and SNBTS to plan the additional support arrangements anticipated following transfer**

- Once the need for transfer is agreed, this should be arranged via Scotstar. The clinician with the patient should call the referral line on 03333 990 222. This allows both the critical care bed to be allocated in the appropriate centre and the transfer to be arranged. A conference call with the local clinician, the retrieval clinician, the accepting critical care consultant and the haematologist will be facilitated via Scotstar

Criteria for Early Transfer
For sickle cell disease:

- **Symptoms or signs suggesting acute chest syndrome/girdle syndrome (links)**
  Early discussion by telephone with the on-call paediatric haematology consultant at the referral centre is recommended to aid preparation of staff / facilities for potential exchange transfusion.

- **Transient ischaemic attack (TIA), seizure and overt stroke or severe acute headache**
  Early discussion by telephone with the on-call paediatric haematology consultant at the referral centre is recommended to arrange prompt transfer for exchange transfusion. This should not delay commencing treatment with top up transfusion therapy if appropriate at the local hospital (link to stroke guideline)

- **Severe sepsis**
  Early discussion by telephone with the on-call paediatric haematology consultant at the referral centre is recommended for any child presenting with severe sepsis to discuss transfer for availability of critical care support

- **Suspected osteomyelitis**
  Prior to orthopaedic intervention, all children with suspected osteomyelitis should be discussed and arrangements made to review imaging if appropriate.

- **Urgent or complex surgery, vascular access and removal**
  These patients should be discussed with the on-call paediatric haematology consultant to liaise with the appropriate surgical team for further management and consideration of need for urgent exchange transfusion prior to the procedure

- **Acute visual impairment**
  Children presenting with acute visual impairment should be discussed with the on-call paediatric haematology consultant and ophthalmology review organised

- **Fulminant Priapism**
  These patients should be discussed with the on-call paediatric haematology consultant to plan for urological intracavernosal aspiration / washout and potential Winter's shunt. The haematology consultant will liaise with urological/ PICU services to plan medical management / surgical intervention and exchange transfusion if necessary. Place of transfer may depend on availability of appropriate surgical expertise

- **Hyperhaemolysis syndrome**
  These patients should be discussed with SNBTS transfusion services in addition to the paediatric haematology consultant to plan appropriate treatment. Discussion with PICU for additional support may be necessary

- **Severe liver dysfunction** including biliary sepsis / hepatic sequestration and hepatitis These patients should be referred for joint haematology / hepatology input following discussion with the on-call paediatric haematology consultant. If necessary arrangements can be made for ERCP / MRCP. Severe liver failure should be discussed with the liver unit at Birmingham via the local hepatology/GI team. If there is a need to transfer the patient to Birmingham

or London for specialist hepatology care the process would be via Scotstar as above.

- **Acute renal impairment**
  These patients should be discussed with the on-call paediatric haematology consultant and transfer considered if appropriate for renal support including haemofiltration filtration. Place of transfer may depend on availability of appropriate renal specialist expertise.

For Thalassaemia:

- Early discussion by telephone with the on-call paediatric haematology referral centre consultant is recommended for any child presenting with severe sepsis
- Chelation associated severe cytopenia, liver dysfunction or renal impairment should be discussed with the on-call paediatric haematology referral centre consultant and transfer organised if appropriate for renal, liver or high dependency input
- Children presenting with acute complications of endocrinopathy (severe hypothyroidism, hypoparathyroidism, diabetes) should be discussed and transfer organised if appropriate

- Children presenting with acute cardiac decompensation should be discussed urgently and arrangements made for critical care transfer to RCH Glasgow via Scotstar.

Any other life-threatening complications requiring urgent critical care input should be discussed with the on-call paediatric haematology Consultant, but transfer should not be delayed.

Acknowledgement:
Thank you to the Haematology Department of Barts Health NHS Trust, London for allowing us to reference this guideline

Approval date: November 2021
Review date: November 2024

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.