## STSTN Monitoring and Annual Review of Children Receiving Regular Blood Transfusions (including sickle cell disease and thalassaemia)

<table>
<thead>
<tr>
<th>Document Detail</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Document Type</strong></td>
</tr>
<tr>
<td><strong>Document name</strong></td>
</tr>
<tr>
<td><strong>Version</strong></td>
</tr>
<tr>
<td><strong>Effective from</strong></td>
</tr>
<tr>
<td><strong>Review date</strong></td>
</tr>
<tr>
<td><strong>Authors</strong></td>
</tr>
<tr>
<td><strong>Superseded documents</strong></td>
</tr>
<tr>
<td><strong>Keywords</strong></td>
</tr>
</tbody>
</table>

### Change History

<table>
<thead>
<tr>
<th>Date</th>
<th>Change details, since approved</th>
<th>Approved by</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

DISCLAIMER: This guideline is for information purposes only and is not intended to inform any individual clinical decisions.

STSTN and its members do not accept any responsibility for outcome of clinical decisions made as a result of reading these guidelines. All guidelines have been peer-reviewed and agreed to be published by the relevant lead consultants in the network.
Background

This guideline covers the monitoring and management of children (receiving regular blood transfusions in daycare) with transfusion-dependent anaemias, and is aimed at the paediatric, haematology and nursing members of the multi-disciplinary team. A separate guideline covers Iron Chelation.

Organisation of Blood Transfusions

- Regular blood transfusions (top up or automated exchange) are given on the Day Unit
- These children will be seen in the transfusion clinic 6-monthly for review
- Patients for blood transfusion are booked in to the Day Unit following discussion with the ward staff and consultants and the clinical nurse specialist if available.
- Subsequent appointments for transfusion will be booked by the staff on the Day Unit according specified frequency.
- On deciding to start transfusions, the child’s hepatitis A, B and C status should be checked
- Vaccination against hepatitis A and B should be started if not immune
- The patient’s extended red cell phenotype must be documented
- Irradiated blood is requested for children who are due to undergo Haematopoietic Stem Cell Transplantation (HSCT) or those who have undergone HSCT.

Indications for Regular Blood Transfusions

- Sickle cell disease
  - Primary stroke prevention in children with abnormal transcranial Doppler scans
  - Secondary stroke prevention
  - Severe episodes of acute chest syndrome unresponsive to hydroxyurea
  - Frequent and disruptive episodes of acute pain unresponsive to hydroxyurea
  - Recurrent severe anaemia due to recurrent splenic sequestration, until splenectomy
- Thalassaemia
  - Failure to thrive
  - Abnormal bone expansion
  - Symptomatic anaemia
  - Frequent sporadic blood transfusions
- Others
  - Severe hereditary spherocytosis
  - Severe pyruvate kinase deficiency
  - Diamond-Blackfan anaemia
  - Other rare inherited haemolytic anaemias

STSTN Monitoring and Annual Review of Children Receiving Regular Blood Transfusions (including sickle cell disease and thalassaemia) August 22 2020

DISCLAIMER: This guideline is for information purposes only and is not intended to inform any individual clinical decisions.

STSTN and its members do not accept any responsibility for outcome of clinical decisions made as a result of reading these guidelines. All guidelines have been peer-reviewed and agreed to be published by the relevant lead consultants in the network.
Monitoring of Children Receiving Regular Blood Transfusions

- The paediatric haemoglobinopathy CNS or community sickle cell nurse should be informed in writing of all children receiving regular blood transfusions. They should be included in the MDT list and individual patients discussed at each MDT.

- Target haematological parameters
  - Each child will have a post-transfusion target haemoglobin and HbS% (sickle only), recorded in the MDT notes.
  - The pre-transfusion haemoglobin target is 90-100g/dl unless otherwise stated.
  - Haematological parameters will be reviewed monthly at the MDT and altered to achieve targets.

- Prior to each transfusion, the following laboratory tests will be performed
  - Full blood count
  - Reticulocyte count
  - Haemoglobin HbS% (and HbC%) – (SCD only)
  - Renal and hepatic function, including ALT
  - Bone chemistry/profile
  - Serum ferritin
  - Urine albumin:creatinine ratio - patients on deferasirox (Exjade) only
  - Vitamin D levels (3 monthly)
  - MRSA swabs/COVID-19 swabs as per local protocol

- Clinical assessment at each transfusion
  - If the child has any symptoms, she/he will be assessed by the ward SHO who will liaise with the sickle cell team.
  - Children will be formally examined every 6 months by a paediatric haematology or paediatric consultant, spleen size is recorded.
  - Weight will be recorded at each transfusion and used to calculate the volume of blood to transfuse.

Annual Review of Regularly Transfused Patients

- Review blood tests will be performed annually on all regularly transfused patients as follows:
  - Routine monthly blood and urine tests as listed above
  - Endocrine tests: Thyroid function tests, Parathyroid hormone levels, Insulin-like growth factor 1
  - Random glucose
  - Serum magnesium, zinc, selenium, copper
  - Cystatin C (if available, otherwise renal profile including creatinine)
  - Viral serology: CMV IgG (if previously negative), Hepatitis A, B and C serology
  - Urine for microscopy and culture and albumin:creatinine ratio

STSTN Monitoring and Annual Review of Children Receiving Regular Blood Transfusions (including sickle cell disease and thalassaemia) August 22 2020

DISCLAIMER: This guideline is for information purposes only and is not intended to inform any individual clinical decisions.

STSTN and its members do not accept any responsibility for outcome of clinical decisions made as a result of reading these guidelines. All guidelines have been peer-reviewed and agreed to be published by the relevant lead consultants in the network.
- Other investigations at annual review
  - Audiometry and ophthalmology review for patients on iron chelation
  - Height
  - ECG
- The results of Annual Review blood tests will be discussed at the MDT meeting.
- The child will be reviewed by a paediatric haematology/paediatric consultant with the results in the transfusion clinic.
- This will include:
  - Full examination
  - Discussion of iron chelation, including adherence, treatment options
  - Discussion of venous access, and assessment of Portacath, if in use
  - Review and update of immunizations (including Pneumovax)
  - Assessment of growth and development
  - Pubertal Tanner staging if appropriate
  - Assessment of school attendance and performance
  - The need for continuing blood transfusions
  - Social, housing and financial status
  - Birth of siblings or other family changes which might make HLA typing possible
  - Discussion of bone marrow transplantation
  - Discussion of transition to adult services if appropriate

**Additional Review of Sickle Cell Disease patients receiving Regular Blood Transfusion**
- The majority of these patients have cerebrovascular disease and the following investigations should be performed regularly, typically every 12 months
  - Neurocognitive assessment – for patients with previous stroke or silent infarcts on MRI. This is normally organised by the paediatric haemoglobinopathy psychologist but may need specific referral if a new patient.
  - Brain MRI/MRA – after the age of seven this is typically performed annually, but in younger children general anaesthesia may be required and this will be requested only if there are new neurological symptoms or evidence of progressive vasculopathy.
  - Transcranial Doppler imaging – usually this will be performed every 6-12 months, although it may not be informative or necessary if there are vessels which cannot be assessed due to severe vasculopathy or an inadequate ultrasound window (confirmed on departmental rather than portable TCD).
  - Review in the Combined paediatric sickle/neurology clinic with the consultant paediatric neurologist
- Indications for regular transfusions in patients for non-neurological reasons should be reviewed regularly, as most will not be transfused indefinitely.

**STSTN Monitoring and Annual Review of Children Receiving Regular Blood Transfusions (including sickle cell disease and thalassaemia) August 22 2020**

DISCLAIMER: This guideline is for information purposes only and is not intended to inform any individual clinical decisions.

STSTN and its members do not accept any responsibility for outcome of clinical decisions made as a result of reading these guidelines. All guidelines have been peer-reviewed and agreed to be published by the relevant lead consultants in the network.
• Assessment of liver and cardiac iron loading. If the ferritin is consistently greater than 2500mcg/l, and the child does not need a General Anaesthetic to perform the scan (usually >7 yrs), quantitative assessment of iron overload should be arranged using T2* cardiac MRI and R2 MRI of the liver (Ferriscan). In a young child who is likely to need a GA, discuss with the specialist centre – GA for Ferriscan is currently only available at ECH. Iron chelation is dealt with in a separate guideline.

Additional Review of Children with Transfusion-Dependent Thalassaemia

• Bone densitometry (DEXA scan) should be requested annually from the age of 10 years.
• T2* cardiac MRI should be requested at the age of 7-8 years. This should be repeated approximately annually depending on the initial result and the chelation history of the patient.
• Oral Glucose Tolerance Test at puberty or >10 years - annual
• Cardiac Review >10 years.
• Endocrine Review >10 years unless problems identified before this. For management of bone fractures see separate guideline.
Contact details:

KCH Paed sickle CNS: 02032994752
KCH Paed sickle consultants: david.rees2@nhs.net subarna.chakravorty@nhs.net
KCH Paed Sickle Consultant: sue.height@nhs.net
GSTT Paed sickle CNS: 0207 188 9432
GST Paed sickle consultants: Baba.Inusa@gstt.nhs.uk maria.pelidis@gstt.nhs.uk
QE Paed sickle sickle CNS: 07741233556
QE Paed sickle consultants: julie.lord@nhs.net aruj.qayum@nhs.net
UHL Paed sickle CNS: 07741233556
UHL/QE Paeds sickle consultants: s.wilkinson6@nhs.net julie.lord@nhs.net
CUH Paeds sickle CNS: 0208 2517229
CUH Paeds sickle consultants: nazmachowdhury@nhs.net

Additional contacts can be found on the STSTN website (www.ststn.co.uk)

Guidelines written by the STSTN adult writing group:

Dr Sue Height
Dr Sarah Wilkinson
Dr Rachel Kesse-Adu
Professor Baba Inusa

STSTN Monitoring and Annual Review of Children Receiving Regular Blood Transfusions (including sickle cell disease and thalassaemia) August 22 2020

DISCLAIMER: This guideline is for information purposes only and is not intended to inform any individual clinical decisions.

STSTN and its members do not accept any responsibility for outcome of clinical decisions made as a result of reading these guidelines. All guidelines have been peer-reviewed and agreed to be published by the relevant lead consultants in the network.