King's College Hospital NHS
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Monitoring and Annual Review of Children Receiving Regular Blood Transfusions (including sickle cell disease and thalassaemia)

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Abstract Page

Background

This guideline covers the monitoring and management of children (receiving blood transfusions on Philip Isaacs ward) with transfusion dependent conditions, and is aimed at the paediatric, haematology and nursing members of the multi-disciplinary team. A separate guideline covers Iron Chelation.

Contents of guideline :

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

Organization of Blood Transfusions

- Regular blood transfusions are given on Philip Isaac's Day Treatment Ward.
- These children will not normally attend Outpatient Clinics.
- Patients for blood transfusion should be booked in to Philip Isaac's ward following discussion with the ward staff, consultants and the clinical nurse specialist.
- Subsequent appointments for transfusion will be booked by the staff on Philip Isaac's ward 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

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
- 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

Monitoring of Children Receiving Regular Blood Transfusions

- The paediatric haemoglobinopathy CNS should be informed in writing of all children receiving regular blood transfusions. She will add them to the MDT list and they will be reviewed on the 2nd Friday of each month at the MDT.
- Target haematological parameters
    - Each child will have a post-transfusion target haemoglobin and HbS% (sickle only), recorded in the MDT notes on EPR.
    - The pretransfusion haemoglobin target is 9-10g/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
    - Renal and hepatic function, including ALT
    - Corrected calcium
    - Serum ferritin
    - Haemoglobin HbS (and C) percentage – (SCD only)
    - Urine albumin:creatinine ratio (patients on deferasirox only)
    - Vitamin D levels
- Clinical assessment at each transfusion
    - If the child has any symptoms, she/he will be assessed by the ward SHO who will liase with the paediatric haematogy specialist registrar
    - Children will be formally examined every 6 months by a paediatric haematology consultant
    - Weight will be recorded at each transfusion and used to calculate the volume of blood transfused.

Annual Review of Regularly Transfused Patients

- Annual review will be initiated by the clinical nurse specialist
- Review blood tests will be performed annually on all regularly transfused patients (order set on EPR).
    - Routine monthly blood tests as listed above
    - Thyroid function tests
    - Parathyroid hormone levels
    - Cystatin C
    - Random glucose
    - Serum magnesium, zinc, selenium, copper
    - Insulin-like growth factor 1
    - CMV IgG (if previously negative)
    - Hepatitis A, B and C serology
    - Urine for microscopy and culture
    - Urine albumin:creatinine ratio
- 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 consultant with the results on Philip Isaac's ward, according to the timetable organized by the clinicalnurse specialist. 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
    - 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 have cerebrovascular disease and the following investigations should be performed regularly, typically every 12 months
  - Neurocognitive assessment – normally organized 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 is required and this will be requested only if there are new neurological symptoms or evidence of progressive vasculopathy.
  - Transcranial Doppler imaging – typically this will be performed every 6-12 months, although it may not be informative or necessary if there are assessable vessels due to vasculopathy or inadequate ultrasound window.
  - Review in the paediatric joint sickle/neurology clinic with the consultant paediatric neurologist (list currently managed by Dr Sue Height)
- Indications for transfusions in non-neurological patients should be reviewed regularly as most will not be transfused indefinitely.
- If the child is older than 7 years, and the ferritin is consistently greater than 2500mg/l, T2* cardiac MRI and R2 MRI of liver should be requested (these can both now be done at KCH).
- Iron chelation is dealt with in a separate guideline

Additional Review of Children with Transfusion-Dependent Thalassaemia

- Bone densitometry 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. This is currently requested by referring to Professor Dudley Pennell at the Royal Brompton Hospital.
- If the ferritin is persistently > 2000mg/l or the T2* MRI suggests significant hepatic iron overload, an R2 liver MRI scan should be organized (at KCH/Maudsley
- Iron chelation is dealt with in a separate guideline
- OGTT >at puberty or > 10 years
- Cardiac Review >10 years
- Endocrine Review >10 years

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