South Thames Sickle Cell & Thalassaemia Network

Guideline for the Long Term Management of Stroke in Paediatric Patients with Sickle Cell Disease

Following a stroke in children with Sickle Cell Disease (SCD) there is a high risk of recurrence if no treatment is offered. Most children in the UK will have regular blood transfusions as secondary stroke prevention and the initial aim is to maintain HbS percentage below 30%. Children require regular monitoring to assess the effectiveness of their blood transfusion in preventing progression of their cerebrovascular disease, as well as monitoring for complications including iron overload.

Separate guidelines exist for the details of management of Acute Stroke, blood transfusions, chelation therapy and Hydroxycarbamide (see STSTN website), and below is a summary of the various aspects of the investigation and monitoring required; this is usually arranged on an outpatient basis, in the clinic and day unit.

Additional Measures:

Following diagnosis of stroke/cerebrovascular disease all patients should have:-

- Overnight oximetry study to assess for Sleep Disordered Breathing and referral to ENT or Respiratory clinic for further management as necessary.
- Cardiac ECHO including assessment for PFO (paradoxical embolus)
- Thrombophilia screen
- Lyme serology/TPHA
- HLA-typing of patient and siblings not affected by Sickle Cell Disease (HbSS/S thalassaemia/HbSC); siblings with Sickle Cell Trait (HbAS) would be possible donors if HLA-matched.
- Further clinical assessment by a Paediatric Neurologist (refer to specialist joint clinics at Evelina or King's College Hospitals. Physiotherapy, Speech and Language Therapy and Occupational Therapy input as needed (usually arranged before discharge from hospital as part of neurorehabilitation)
- Ophthalmology assessment (visual fields/acuity etc) for stroke involving the visual pathway.
- Neurocognitive Assessment by Clinical Psychologist
- Involvement of Local Authority, CAMHS and Community Paediatric Services as additional support may be needed in school

Regular Blood Transfusions

Following a stroke regular transfusions are arranged, continuing until adulthood, to reduce the risk of further events:

- The target pre-transfusion HbS% <30%, and may be achieved by regular top-up transfusion or automated exchange transfusion (see separate guideline on transfusion)
- Monitor for iron overload and start chelation therapy when ferritin >1000 or after 1 year of transfusion (see regular monitoring of children receiving iron chelation)
- Discuss venous access and option of Portacath
- After 3 years the HbS% may be allowed to increase to <50% in individual patients where monitoring has not shown progression of cerebrovascular disease, continuing surveillance.
- Children who cannot have regular transfusion (eg multiple alloantibodies) should be offered Hydroxyurea at Maximum Tolerated Dose (MTD), as secondary stroke prevention.

Monitoring

- Annual review by Paediatric Neurologist (suggested interval annually but individual frequency will vary)
- Regular TCDs (6 monthly suggested) unless they are uninformative.
- Regular MRI/MRAs including carotid and vertebral arteries to assess for progression – the frequency may vary between individuals but annual scans are suggested.Scans should be reviewed in a Neuroradiology MDM with the team and paediatric neurologist.
- Ferriscan and Cardiac T2* MRI - annually after starting chelation (if not needing GA)
- Regular neurocognitive re-assessment is on an individual basis n discussion with neuro psychologist.

Progressive Cerebrovascular Disease

For patients where there is evidence of progression on serial imaging or neurocognitive assessments or new events occur despite optimal transfusions, consider

- Intensifying transfusions to achieve HbS<20%
- Adding Hydroxyurea
- Referral for discussion of Revascularisation surgery (Mr Sanj Bassi and Mr Christos Tolias, Neurosurgeons at King's College Hospital) or to the Revascularisation clinic at Great Ormond Street (Evelina patients).
- Consider Stem Cell Transplantation (Discussion in an MDM if no HLA-identical siblings available as donors).If MRI/MRA identifies an aneurysm which is asymptomatic then the patient should be discussed in the Neurovascular MDM at Kings (referral to MDT Coordinator by email: kch-tr.nv-abi-clinics@nhs.net).
-) which takes place every Tuesday 13.00 in the Neuroradiology reporting room.

Stem Cell Transplantation

If a child has an HLA-matched sibling unaffected by Sickle Cell Disease, allogeneic stem cell transplantation should be discussed as a possible option for those who would otherwise require long term transfusion, and a referral made to the Transplant Team at Imperial College for assessment.

For some patients with progressive disease and no HLA-matched sibling donor, families may ask about gene therapy (not yet available in the UK) or haplo-identical transplants – this is experimental treatment at present and should be discussed with the team and in an MDM on an individual basis before referral.

Silent cerebral infarcts (silent strokes)

These are found frequently on MRI/MRA and the options of Hydroxycarbamide and/or transfusion can be discussed. For patients with the following, regular blood transfusion may be appropriate and should be discussed:

- Impaired cognitive function and progressive deterioration
- Progression of changes on serial MRIs
- Evidence of intracranial or extracranial Vasculopathy on MRA
- Other co-existent morbidities eg conditional TCDs, renal impairment