Clinical Guidance

***Long-Term Management of Stroke in Paediatric Patients with Sickle Cell Disease***

Summary

*Stroke in sickle cell disease, acute emergency management as well as follow on management and investigations.*

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| Change History | | |
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**Further Management of Stroke in Paediatric Patients with Sickle Cell Disease**

## This guideline follows the treatment of acute stroke event in a child with sickle cell disease, it recommends additional investigations essential in future monitoring:

## **Re-View the following**

* CT Scan
* Arrange urgent neuro-imaging- MRI with dWI / MRA and angiographic sequences. If symptoms suggest posterior circulation involvement consider request for fat suppressed sequences of neck vessels
* Consider urine and serum drug screen if altered mental status with no explanation.
* Consider EEG if marked unexplained encephalopathy
* MRI/MRA including neck vessels and perfusion-weighted images discuss with neuroradiology; Children <6 years may require GA and the ward paediatric staff will need to contact the on-call anaesthetist.
* TCDs including extracranial ICAs
* Sleep Study
* Trans-thoracic cardiac echo (discuss with Paediatric Cardiology Team) Further investigations may be needed to exclude a Patent Foramen Ovale – bubble studies or trans-oesophageal echo (under GA)

## **Further Investigations**

* All patients must have:
* Sleep study e.g. home oximetry studies to rule within 4 weeks to assess the degree of hypoxia
* Cardiac evaluation -ECHO including assessment for PFO which may account for thromboembolic phenomenon
* Fasting blood sugar?
* Cervical MRI to exclude external carotid thrombus
* Lupus anticoagulant Immunoglobulins assay, auto-antibodies, ANCA, full thrombophilia screen including anti-Cardiolipin antibodies. (Anti-thrombin, free Protein S, Protein C, APCR, FV Leiden, Prothrombin 20210A mutation, lupus anticoagulant screen, MTHFR 677), anti-cardiolipin antibodies, homocysteine, lipoprotein-a, cholesterol, PNH screen
* TPHA/Lyme serology

Clinical Evaluation of all patients for the history of prior infection (varicella), immunisation, dysmorphic features, neurocutaneous

# Neurorehabilitation and Subsequent Management / referral on discharge

* Institute neurorehabilitation before discharge
* Speech/physiotherapy as necessary
* Ensure daily assessment by the Neurology Registrar within the first 7days
* Organise Neuro-psychology assessment before discharge.
* Refer to Joint Sickle/Neurology Clinic
* Arrange regular blood transfusion with target HbS <30%, 3-5 weekly for top up transfusion and 6-8 weekly for exchange transfusion programme.

**Principles of Regular Blood Transfusions**

* Following a stroke, children are transfused regularly into adulthood to prevent the occurrence of further strokes
* Aim for a target pre-transfusion HbS% < 30%
* After 3 years, of consistent transfusion, the HbS% may be allowed to rise to <50%
* Children who cannot receive regular blood transfusion might be considered for hydroxyurea
* Monitor ferritin and discuss iron chelation therapy (to commence when ferritin >1000), baseline liver FerriScan at onset of chelation
* Monitoring for iron overload (see guidelines)

# Long Term Management

* Yearly joint clinic with paediatric neurologist
* Yearly MRI including MRA scans to rule out any progression of Cerebrovascular changes
* Annual Transcranial Doppler Scans (TCD) including assessment of extracranial vascular changes.
* Regular Neurocognitive testing
* Hydroxycarbamide should be considered as part of secondary prevention when blood is not suitable e.g. multiple antibodies, or as alternative where blood transfusion is not acceptable to the family
* Consider anticoagulant therapy in presence of other risk factors
* Consider haematopoietic stem cell transplantation in children and young people staring on regular blood transfusion

**Silent Cerebral Infarct Lesions in SCD**

* Silent Cerebral Infarction- Discuss the benefit of blood transfusion with the children, families and neurologists. Factors favouring transfusion:
  + Impaired cognition
  + Increasing size and / increase numbers of silent cerebral infarct lesions
  + Other co-existing morbidities of SCD including pain, conditional TCD, progressive renal impairment.

Ref

RCPCH 2017 stroke guidelines