

**GSTFT Clinical Practice Guideline**

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

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<b>Authors:</b>	<b>Dr Su Li, Dr N Boggis, Dr Baba Inusa</b>
<b>Speciality:</b>	<b>General Paediatrics &amp; Haemoglobinopathies</b>
<b>Directorate:</b>	<b>Children's and Genetics</b>
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## **Following treatment of the acute event, further investigations should be undertaken as appropriate:**

### **Further Investigations to be considered**

Overnight O<sub>2</sub> saturation monitoring

ENT assessment

Consider X-ray cervical spine if appropriate

Immunoglobulins/ANCA

Thrombophilia screen & thrombophilia risk factors (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

### **Subsequent Management and referral on discharge**

Daily Neurological Assessment within the first 7days

Neuro-psychology assessment

Speech/physiotherapy as necessary

Refer to Joint Sickle/Neurology Clinic

Arrange regular blood transfusion with target HbS <30%

### **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)

Monitoring for iron overload and desferal toxicity (see guidelines)

Monitoring of vasculopathy

Annual transcranial dopplers

Periodic neuro-imaging

Neuro-psychometric assessment