South Thames Sickle Cell & Thalassaemia Network

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

Summary
Outline of the presentation of stroke in paediatric patients with sickle cell disease, urgent investigations, emergency management and arrangements for referral and transfer.

Definition of Stroke and TIA in Paediatric Patients with Sickle Cell Disease
Cerebrovascular Accident (CVA) is a neurological event lasting >24 hours with radiographic evidence of new areas of abnormality.

Transient Ischaemic Attack (TIA) is a focal event lasting < 24 hours with no radiographic evidence of abnormality.

Stroke in Paediatric Sickle cell disease:

- Clinical stroke is 250 times more common in children with Sickle Cell Disease (SCD) than the general paediatric population
- There are two main types of stroke: infarction resulting from arterial occlusion, and haemorrhagic due to an intracranial bleed. In childhood, the majority of strokes in SCD are due to infarction.
- Before Transcranial Doppler Scan (TCD) screening to identify those at increased risk, 11% of children with SCD had an overt stroke by the age of 16 (peak age 7) and the incidence is highest in the first decade.
- The rates of CVA vary by sickle genotype. The age adjusted incidence of CVA is highest for those with HbSS (0.61/100 person-years), and HbSβ^0 thalassemia compared with HbSC (0.15/100 person-years) or HbSβ^+ thalassaemia (0.09/100 person-years).
- Haemorrhagic strokes are more common in the second decade
- 10-25% of asymptomatic children with SCD have an abnormal MRI showing silent cerebral infarcts.

Presentation:

- There is a wide spectrum of presentation of stroke in children with SCD.
- Common presentations of an acute ischaemic stroke include motor deficits such as hemiparesis, monoparesis, aphasia or seizure. Posterior circulation strokes may present with ataxia, headaches, vertigo or vomiting

- In young children symptoms may be subtle and mistaken for another illness
- Haemorrhagic stroke may present with acute, severe headache
- TIAs may present with features similar to an ischaemic stroke but resolve spontaneously
- A high index of suspicion is required. In children with SCD any new seizures, changes in personality, inability to move limbs and other subtle changes in behaviour including communication should alert you to the possibility of stroke.
- Consider other diagnoses (see Page 4) whilst proceeding with urgent management of possible stroke.

If stroke is suspected in a child with sickle cell disease follow the protocol as below:

Suspected Stroke

- Acute onset focal neurological deficit e.g. facial asymmetry, hemiparesis,
- New onset focal seizure
- Speech disturbance
- Altered conscious level
- New onset severe headache
- Ataxia
- **History of any of the above but signs now resolved (possible TIA)**

Emergency Management at the Hospital where the child presents acutely

- Manage Airway, breathing, monitor SpO2 and maintain oxygen saturations >96%
- Secure IV access and take urgent blood samples (see below)
- 0.9 % Normal saline maintenance IV fluid or two thirds maintenance (avoid fluid overload)
- Manage hypoglycaemia
- Control seizures
- Assess GCS
- **Arrange urgent imaging within 1 hour of arrival: CT head** can be performed without GA/sedation (identify acute bleed/space occupying lesion)
- Admit to HDU/PICU or contact specialist centre (see below for contact information) to arrange urgent transfer.
- Plan for urgent top up transfusion (see below) if any delay in transfer/Hb <80g/l)
- See Differential Diagnosis table below for other things to consider in assessment- depending on clinical presentation it may be necessary to add broad spectrum antibiotics with CNS penetration, with IV Aciclovir to cover for possible intracranial infection and consider LP.

Urgent Blood tests

Haematology

- FBC, Reticulocytes, film, HbS% and HbF%
- PT/APTT and Clauss Fibrinogen
- Blood Group (ABO RhD and Kell & antibody screen – extended red cell phenotype if not previously documented) and urgent cross match (request sickle negative blood)

Biochemistry

- Blood glucose
- Blood gas analysis - Venous (Arterial if arterial line available)
- CRP, Urea and Electrolytes, Calcium, Magnesium
- Liver Function tests, ALT and LDH

Infection screen:

Monitoring

- HR/RR/BP/SpO2
- Neurological Observations
- Blood Glucose and Fluid balance
- Inform paediatric Nurse Practitioner (PNP), Paediatric SpR, PICU and paediatric/paediatric haematology consultant on-call (depending on hospital)

Contact a Sickle Cell Centre to arrange urgent transfer (Evelina, King's or St George's) where urgent exchange transfusion and further imaging can be performed.
Depending on the patient's clinical condition and STRS retrieval team may be required for the transfer.

If an intracranial haemorrhage or space occupying lesion is identified on CT the neurosurgeons (KCH or St George's) should be contacted directly and they can view the images; if the patient is transferred for neurosurgical or interventional neuroradiology treatment, the sickle cell team at the centre will need to be informed as well to arrange exchange transfusion.

Patients will usually be transferred to T&G ward but if an HDU/PICU bed is needed this will be discussed.

Further imaging can then be arranged – MRI/MRA can be arranged at Sickle Cell Centre after transfer – may required GA for young children.

- MRI/MRA with Diffusion weighted images, including head & extracranial carotid & vertebral arteries

Simple top up transfusion while preparing for Exchange blood transfusion *(Hb>70g/L) we would top up at higher Hbs*

Blood Transfusion:
Patients with suspected or confirmed CVA will need an exchange blood transfusion aiming for target HbS<30% and Hb ≤120g/l and this will be done at the Specialist Centre after transfer; it may be an automated or manual exchange. Anaemia must be corrected first by simple top up transfusion to Hb 100g/l if there is any delay in transfer and /or Hb < 80g/l.
See separate exchange protocol for details of procedure.

Differential Diagnoses to consider	**Symptoms/Signs**
Meningitis/encephalitis	Severe headache, neck stiffness, photophobia Rash, fever Altered behaviour
Syncope	Sudden Loss of Consciousness without fit? Vasovagal/cardiac
Stroke	Altered mental state Aphasia, hemiparesis, ataxia, vertigo, coma
TIA	Acute deficit resolves <24 hours and normal neuro imaging
Sub- arachnoid Haemorrhage	Severe headache/neck stiffness +/- deficit
Vaso-occlusion of calvarium	Headache with tenderness +/- scalp oedema
Cerebral Malaria	Altered conscious level, background history of travel to malaria prone area
Trauma	Fractures/contusions
Fat embolism	Severe painful episode, desaturation, coma, petechial rash, multi-organ failure, DIC
Drugs	Altered mental state and other related to agent Enquire about: opiates, paracetamol, NSAIDs, alcohol, non-prescribed drug use.

Abscess	Headache, fevers, Focal signs Background of sinusitis, otitis, mastoiditis?
Tumour	Headache, progressive focal signs, papilloedema
PRES: (Posterior Reversible Encephalopathy Syndrome)	Hypertension, acute visual loss, seizures, recent Acute Chest Syndrome
Cerebral Venous Sinus Thrombosis (CVST)	Dehydration, inflammatory disorders, OCP, intracranial sepsis – MRV imaging

Following Transfer to Specialist Centre

Stabilise as above and arrange urgent investigations (including baseline tests not performed at local hospital:

- Request Blood for Exchange Transfusion (see separate protocol)
- Venous/arterial lines – HDU/PICU as required
- Arrange MRI MRA (head and carotid/vertebral arteries) with diffusion weighted imaging – for children <7 years this may require GA and should only be done after exchange and when the patient is stable.
- Consider MRV if possible cerebral venous sinus thrombosis
- Transcranial Doppler including with extracranial vessels
- Inform Paediatric neurology team and arrange review within 24 hours of admission
- Physiotherapy, Speech & Language Therapy, Occupational Therapy referrals
- Neuro-psychometric Assessment – referral to Clinical Psychologist with Sickle Cell Team.- see further management protocol for details
- Cardiac Echo – to exclude embolic cause for CVA
- Sleep Study

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Contact details:
If you have any questions or concerns about issues raised in this guidance, your medicines, or other queries on your health, please speak to the staff caring for you in your local centre.

Each team to please insert your centres contact details here:

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

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