South East London and South East

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

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Summary

This guideline provides an overview of the presentation and emergency management of stroke in paediatric patients with sickle cell disease. The priority is emergency transfer to a specialist sickle cell centre with a paediatric intensive care unit and arrangement for an exchange transfusion. This guideline outlines the urgent investigations, emergency management, including top up blood transfusion at the local hospital if any delays to transfer anticipated, and arrangements for referral and transfer to an specialist centre.

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). 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 third 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.

Differential Diagnosis of Acute Neurological Presentations in Sickle Cell Disease

Differential Diagnoses to consider	**Symptoms/Signs**
Meningitis/encephalitis	Severe headache, neck stiffness, photophobia Rash, fever Altered behaviour
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
Syncope	Sudden Loss of Consciousness usually without fit Vasovagal/cardiac
Trauma	Fractures/contusions
Seizure	Altered consciousness, tonic/clonic or absence, incontinence, tongue bitten. Post-ictal drowsiness
Drugs	Altered mental state and other related to agent Enquire about: opiates, paracetamol, NSAIDs, alcohol, non-prescribed drug use.
Fat embolism	Severe painful episode, desaturation, coma, petechial rash, multi-organ failure, DIC
Abscess	Headache, fevers, focal signs Background of sinusitis, otitis, mastoiditis
Cerebral Malaria	Altered conscious level, background history of travel to malaria prone area
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

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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 resolved within 24 hours (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 considerations 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:

- Blood culture, urine, throat swab and ASO titres
- Viral serology: HSV, CMV, Varicella ZV, Parvovirus, Hepatitis A, B, and C serology
- Malaria screen if foreign travel

Immunology:

- Autoantibody screen with dsDNA antibodies
- Anti-cardiolipin and β2 glycoprotein antibodies

Other

- Consider urine/serum drug screen if altered mental status with no explanation
- Check – most recent TCD result (all children with HbSS /HbSb0thal should have TCD scans annually from age 2-16 years)

Monitoring

- HR/RR/BP/SpO2
- Control of high blood pressure is important, but a rapid fall in blood pressure risks worsening ischaemic/haemorrhagic damage
- 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 the urgency to reach one of these centres for red cell exchange, The South Thames Retrieval Service (STRS) retrieval team may be required for the transfer. While waiting for transfer, consider top up transfusion locally.

See also Pan London Guideline for the Management of Acutely Unwell Children with Sickle Cell Disease- this is a guidance that specifically covers the process of escalating care of critically unwell children with SCD to a SHT with co-located PICU 2022-1006-SOP-Paediatric-Sickle-Cell-Disease-London-Escalation-Guidance-Final-V1.4.pdf (cats.nhs.uk)

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

Contact information
Evelina Children's Hospital Switchboard 020 7188 7188
Monday–Friday 09.00-17.00: Paediatric Sickle Cell Nurses bleep 2733
Monday-Friday 17.00-09.00, Bank Holidays and Weekends Haematology Registrar through switchboard bleep 0294 Number with answerphone – 020 7188 9432

King's College Hospital Switchboard 020 3299 9000
Monday-Friday 09.00-17.00: Paediatric Haematology SpR (via Switchboard 020 3299 9000 and ext 39823) or Paediatric Haematology Consultant on Attending rota (via switchboard/Rotawatch)
Monday-Friday 17.00-09.00 or Bank Holidays/Weekends: Haematology SpR on call (2nd on call during day at weekends, bleep 544) via switchboard, or Red Cell Consultant Haematologist via Switchboard.
Patients will usually be transferred to T&G ward but if an HDU/PICU bed is needed this will be discussed.

St George's Hospital Switchboard 020 8672 1255
Monday-Friday 09-00-17.00: paediatric haematology SpR bleep 7080 or paediatric haematology consultant on call.
Monday-Friday 17.00-0900, Weekends, Bank Holidays: haematology SpR on call (bleep 6068 until 21.00 then mobile via switch) or paediatric haematology consultant on call

Blood Transfusion:
Patients with suspected or confirmed CVA will need an urgent exchange 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. See separate exchange protocol for details of procedure.
Anaemia must be corrected first by simple top up transfusion to a maximum Hb of 100g/l if there is any delay in transfer and/or Hb < 80g/l. This should be considered locally whilst awaiting/coordinating the transfer to specialist centre.

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
- Red cell exchange should be performed as soon as is practically possible, ideally within 6 hours of presentation. Manual exchange should be performed if the child is <30kg, or there would be an unacceptable delay for an automated exchange to be performed
- If the stroke event is haemorrhagic, red cell exchange is less urgent and should be done in collaboration with any plans for neurosurgical intervention
- 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 cerebral venous sinus thrombosis remains in the differential diagnosis
- 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

NB: there is no standard role for thrombolysis using tPA nor for anti-platelet therapy in the management of sickle cell related ischaemic stroke events.

Contact details:

KCH Paed sickle CNS: 020 3299 4752
KCH Paed sickle consultants: david.rees2@nhs.net; subarna.chakravorty@nhs.net
sue.height@nhs.net; john.brewin@nhs.net
KCH paediatric haematology SPR: mobile extension 39283
GSTT Paed sickle CNS: 020 7188 9432
GST Paed sickle consultants: samah.babiker@gstt,nhs.uk; Nick.Fordham@gstt.nhs.uk
GSTT paediatric haematology SPR: Bleep 1621
QEW Paed sickle CNS: 07741233556
QEW Paed sickle consultants: a.sobande@nhs.net aruj.qayum@nhs.net
UHL Paed sickle CNS: 07741233556
UHL Paed sickle consultants: s.wilkinson6@nhs.net
CUH Paeds sickle CNS: 0208 2517229
CUH Paeds sickle consultants: nazmachowdhury@nhs.net

Additional contacts can be found on our website: www.selsehcc.co.uk

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