

GSTFT Clinical Practice Guideline

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

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Cerebrovascular Accident (CVA) is a neurological event lasting >24 hours +/- 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 SCD than the general paediatric population
- 11 % of children with SCD will have an overt stroke by the age of 16 years (peak age 7 years)
- 10 - 25 % of asymptomatic children have an abnormal MRI compatible with the diagnosis of 'silent cerebral infarct',
- Typical abnormalities shown on brain imaging in paediatric patients with SCD and stroke include occlusion or stenosis of the proximal middle cerebral artery (MCA) or distal internal carotid artery (ICA).

Presentation

There is a wide spectrum of clinical presentations of stroke in SCD. Patients may present with a clear acute neurological event, or with more subtle changes e.g. a painless limp, speech problems, personality change or headache, when the diagnosis might be missed unless it is considered.

History and Examination

In combination with usual history ask about:

- Weakness
- Speech difficulties
- Change in personality
- Drugs (prescribed and non-prescribed).

Examination should include:

- Full systems examinations (Neuro/RS/CVS/GIT/MSK/ENT)
- Assessment of GCS
- Assessment of neck stiffness/papilloedema
- Full observations

Initial Investigations & Management

- Admit to Mountain Ward or PICU for exchange transfusion
- Inform Paediatric PNP / SpR & PICU SpR
- Inform Paediatric Neurology SpR immediately and ensure review within 24 hours of admission
- Assess & secure airway – discuss with PICU if decreased consciousness
- O₂ therapy
- Obtain IV access and send blood samples (see below)
- Check blood glucose
- Start IV fluids (75% maintenance)

If febrile

- Urinalysis
- Blood, urine and throat swab for cultures and ASO titre
- Consider LP to rule out meningitis / encephalitis
- Consider malaria screen especially with recent history of foreign travel
- Treat aggressively with broad Ceftriaxone 80mg / Kg daily & macrolide-Clarithromycin 7.5mg / kg 12 hourly

Continue to monitoring

- GCS / neurological observations
- BP / HR
- RR / O₂ saturations
- Arrange urgent neuro-imaging – MRI or CT (if MRI not available urgently)
- Consider urine & serum drug screen if altered mental status with no explanation
- Perform exchange transfusion, target HBS < 30%. If neurological event has occurred on background of severe anaemia (e.g. splenic sequestration or aplastic crisis),

Differential diagnosis of Acute Neurological Problems in Sickle Cell Disease

Diagnosis	Symptoms
Meningitis/encephalitis	Severe headache/fever/neck stiffness/photophobia
Syncope	Sudden LOC without fit? Vasovagal/cardiac cause
Stroke	Altered mental state/coma/aphasia/ataxia/vertigo
TIA	Acute deficit resolves < 24 hours a normal neuro-imaging
SAH	Severe headache/stiff neck +/- def
Vaso-occlusion of calvarium	Headache with tenderness +/- sca oedema
Cerebral Malaria	Recent history of travel to malaria endemic areas
Trauma	
Fat embolism	Severe painful episode, desaturati coma, petechial rash, liver/renal/respiratory failure, rhabdomyolysis, DIC
Drugs	Opiates/aspirin/paracetamol/alcohol
Abscess	Headache/fever/focal signs ? sinusitis/otitis/mastoiditis
Tumour	Headache/ progressive focal signs papilloedema

Blood Tests		Ordered
Haematology	Blood group & screen	
	Crossmatch – See exchange transfusion protocol	
	FBC	
	Blood Film	
	Reticulocytes	
	Haemoglobin analysis – HBS% & HBF%	
	Clotting screen	
Biochemistry	Urea & Electrolytes inc creatinine	
	Liver function tests	
	Bone profile	
	CRP	
	Glucose	
	LDH	
	Serum ACE	
Infection	Blood culture	
	ASOT	
	HSV serology	
	CMV serology	
	Varicella serology	
	Parvovirus serology	
	Hepatitis A, B & C serology	
Immunology	Auto-antibodies & ds DNA antibodies	

Subsequent investigations to be booked

- 24 hr ECG / Cardiac ECHO in discussion with cardiology
- MRI/MRA
- Arrange Transcranial Doppler scanning with Dr Padayachee-86778

Paediatric Sickle Cell Contacts

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