1.0 Introduction

Stroke occurs in 5–10% of people with Sickle Cell Disease. The risk of stroke is highest in such children between 1 and 9 years of age. Arterial ischemic strokes are more common in children, whereas hemorrhagic strokes occur more frequently in adults (ages 20–29). Children with SCD are at increased risk of having an underlying cerebral arteriopathy pre-disposing them to transient ischemic attacks (TIAs), recurrent arterial ischemic strokes and cerebral hypoperfusion injuries.

Thrombosis and intimal hyperplasia, the precursors of ischemic stroke, are thought to result from a combination of factors seen in Sickle Cell Disease. These include high blood-flow velocity in cerebral vessels, rigidity of circulating RBCs, adherence of RBCs to vessel walls, and intravascular sludging. Stroke occurs when the narrowing is severe enough to compromise distal flow, or a thrombus dislodges and causes distal embolization. Hemorrhagic strokes are thought to result from rupture of fragile vessels, although mechanism is not often clear. The risk of ischemic strokes correlates with severity of disease, previous stroke, silent infarction on MRI, sickling with history of stroke, HbS concentration, severity of anaemia, and elevated transcranial doppler (TCD) velocity. Without treatment, 1/3 of patients with CVA will have recurrent strokes, usually within 3 years. The recurrence rate is reduced significantly by a chronic transfusion program (maintaining a level of HbS <30%).

Target Users:

- Clinicians managing patients with Sickle Cell Disease who present acutely with a change in neurological status in the emergency department, in-patient wards and the critical care units.

Target population:

- Children with Sickle Cell Disease who have an acute change in neurological status.

Clinical Features

- **Arterial Ischemic stroke** typically presents acutely with signs and symptoms of hemiparesis or hemisensory, severe/thunderclap headache, visual impairment, visual field deficits, aphasia, ataxia, dysarthria, cranial nerve palsies, or acute change in level of consciousness and sometimes seizures.
- **Hemorrhagic strokes** usually present with more catastrophic generalized phenomena such as coma, headaches, and seizures.
- **Transient ischemic attacks (TIA)** are defined by neurological signs that resolve within 24–48 hours; they are often a precursor to arterial ischemic stroke and should be treated as an emergency.
Note: Treat all patients with appropriate analgesics and antipyretics as per Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease and Fever: Guidelines for Management in Children with Sickle Cell Disease. Refer to the e-formulary for additional information.

Recommendations for Emergency Department Treatment

**Arterial Ischemic Stroke: Guidelines for ED Management in Children with Sickle Cell Disease**

- **Child with Sickle Cell Disease presents with suspected Stroke in ED**
  - Provide immediate assessment and management including:
    - Stabilize vital signs
    - Provide life support if indicated
    - Administer oxygen to maintain $O_2$ saturation $\geq 95\%$
    - Treat seizures and increased intracranial pressure if indicated

- **Initiate stroke protocol in Hyperacute Arterial Ischemic Stroke Pathway** (refer to ED Hyperacute Arterial Ischemic Stroke Epic order set)
  - Note: TPA is contraindicated in patients with stroke secondary to sickle cell

- **Consult**
  - Haematology fellow and Acute Care Neurology Team (M-F, 0900-1700hrs) or Neurology on call (holidays, weekends and after hours) and notify of a Stroke Alert
  - Haematology fellow should see all sickle cell patients with stroke and discuss with staff

- **Keep NPO and establish IV fluid maintenance**
  - Add IV fluids (use Normal Saline unless glucose $< 55\mathrm{mg/dL})$
  - Serum electrolytes should be ordered prior to IV fluid administration as per the Fluid and Electrolyte Administration in Children recommendations

- **Complete Diagnostic Imaging URGENTLY**
  - Notify Neuroradiology
  - Notify Anaesthesia (if needed)
  - MRI/MRA is very sensitive in detecting intracranial haemorrhage or infarction
  - Request CT scan (without contrast) if MRI is contraindicated

- **Other tests:**
  - CBC, diff, reticulocyte count, electrolytes, magnesium, calcium, phosphate, blood typing and cross-matching (ensure Sickle Cell Disease is written on requisition),
  - Blood and urine cultures if patient is febrile; and
  - Blood for coagulation screen (INR, aPTT), fibrinogen and D-dimers.

- **End of ED Management Recommendations**
  - Exchange transfusion will take place in PICU

**PICU and Inpatient Recommendations on next page**

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5.0 Recommendations for In-patient Management: PICU and Ward

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Admit to PICU for exchange transfusion

If the child is febrile, refer to: Fever Guidelines for Management in Children with Sickle Cell Disease and Consult Infectious Disease

Continue IV fluids at max maintenance flow rates

Total fluid intake not to exceed maintenance

For diagnosed arterial ischemic stroke, and/or clear history/physical indicating stroke: perform double-volume RBC exchange transfusion to a haemoglobin of 100g/L and HbS level of <30% of total Hgb (see Exchange Transfusion Protocol, in attachments).

If Hb > 70g/L, commence transfusion of pRBC, while awaiting exchange transfusion; and if patient <20Kg, add 250 cc pRBC to prime circuit (Sickle Cell screened). Refer to Red Cell Exchange/Depletion Order Set.

Order Pre-exchange transfusion labs: CBC, diff, Hb electrophoresis, ionized Ca, K, Mg, Phos, TCO₂

Call blood bank

Remove the central venous line as soon as possible after the blood exchange to reduce the risk of thrombosis

Order ECG and ECHO with bubble study.

Complete as soon as possible (within 24 hours) to assess for intracardiac shunt, thrombus, vegetation.

End of PICU Management

Pediatric Medicine Discharge Preparedness

Encourage ambulation and activity (consult with PT/OT)

(Hospital Childlife representative can recommend structured daily activity)

Inform Sickle Cell Team

Organize clinic follow-up and next transfusion

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References


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**Attachments:**

- Exchange Transfusion Calculation.pdf
- Revision History.docx
- sickle cell_ED_Pathway_2021_Final.pdf
- sickle cell_inpatient_Pathway_2021_FINAL.pdf