IWK Health	Document Scope: Hospital-wide Patient Care	
	Document Type: Clinical Practice Guideline Approved on: January 2024 Next Review Date: January 2027	
	Stroke: Guidelines for Management in Children with Sickle Cell Disease (Adapted from the Hospital for Sick Children)	Version: 1

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 anemia, 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 hemi- anesthesia, 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 Sickle Cell Pain Admission Orders **Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease**

2.0 Stroke: Guidelines for Management in Children with Sickle Cell Disease

Recommendations for Emergency Department treatment

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 greater than 93%.
- Treat seizures and increased intracranial pressure if indicated.
- Call IWK PICU directly if intensive care required.

Initiate Stroke Management

Note: TPA is contraindicated in patients with stroke secondary to Sickle Cell

Gather history and complete physical exam

Acute Stroke Neuroprotective Care

- **NPO:** head of bed flat (if tolerated and no signs of increased ICP)
- **Normotension**: aim for SBP 50-90%ile for age to maintain cerebral perfusion pressure. Tx hypotension (25%ile for age): NS, inotropes. Tx hypertension (>33% above the 95th percentile for age) lower by approximately 25% over 24 hours.
- **Normovolemia**: maintenance normal saline, bolus PRN;
- **Normal O_2, CO_2, pH**;
- **Normothermia**: Treat >37.5°C with antipyretics +/- cooling;
- **Normoglycemia**: No IV glucose unless hypoglycemic, target 5-10mmol/L; and
- **Seizure control**: ASAP with any suspected seizure activity. Consider EEG to monitor subclinical seizures.

History to include:

- quality, timing, severity, and duration of headaches;
- previous headaches;
- nausea or vomiting;
- drooling; visual changes;
- paresis;
- loss of coordination;
- parasthesias;
- fever;
- syncope;
- seizures;
- previous stroke or TIA history;
- recreational or prescribed drug use; and
- result of most recent TCD.

Physical exam to include:

- vital signs;
- detailed neurologic exam;
- hydration status;
- spleen size;
- baseline haemoglobin;
- presence of jaundice; and
- signs of infection

Consult:
Hematology/Oncology and Neurology and prepare to transfer to IWK.

Keep NPO and establish IV fluid maintenance.
*add IV fluids (Use Normal Saline unless glucose < 5 mmol/L)
(Serum electrolytes should be ordered prior to IV fluid administration)

Complete **Diagnostic imaging** URGENTLY

- Notify Neuroradiology.
- Notify Anaesthesia (if needed).
- ED or hematology/oncology to request either CT or MRI as below.
- MRI/MRA is very sensitive in detecting intracranial haemorrhage or infarction.
- Request CT scan (without contrast) if MRI is contraindicated.

Note: CT scan during ED visit may appear normal; CT scan conducted 2-7 days post CVA usually shows areas of infarction.

Other tests:

- CBCD, reticulocyte count, electrolytes, magnesium, calcium, phosphate, type, and screen and cross-match, blood and urine cultures if patient is febrile; and
- Blood for coagulation screen (INR, aPTT), fibrinogen and D-dimers.

Note: If the child is febrile, refer to IWK or APPHON Sickle Cell Fever Orders (e-access or apphon-rohppa.com)

If moderate to severe **pain**, refer to the Sickle Cell Pain Admission Orders and consult Acute Pain Service for hard to manage pain.

Begin the exchange transfusion preparation.
Exchange transfusion will take place in PICU.

End of ED Management Recommendations

PICU and Inpatient Recommendations on next page

Recommendations for Inpatient Management: PICU and Ward

Admit to PICU for exchange transfusion

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If the child is febrile, refer to: **Fever Guidelines for Management in Children with Sickle Cell Disease** on apphon-rohppa.com or IWK Access E-Forms.

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Continue IV fluids

Total fluid intake not to exceed maintenance

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For diagnosed arterial ischemic stroke, and/or clear history/physical indicating stroke: perform double-volume RBC exchange transfusion to a hemoglobin of 100g/L and HbS level of <30% of total Hgb.

If patient < 20 kg add 350 mL pRBC to prime apheresis machine (sickle cell screened). If Hb < 90 g/L commence transfusion of pRBC, while awaiting exchange transfusion (to calculate transfusion requirements go to the app RBCX Calculation).

Order Pre-exchange transfusion labs: CBCD, Hb electrophoresis, ionized Ca, K, Mg, Phos, TCO_2

Call blood bank

Remove the central venous lines as soon as possible after the blood exchange, to reduce the risk of thrombosis.

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Order ECG and **ECHO** with bubble study.

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

End of PICU Management

Pediatric Medicine Discharge Preparedness

Encourage mobility and activity (consult PT/OT for complex needs)

(Child Life Specialist can recommend structured daily activity)

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Inform Sickle Cell Team

Organize clinic follow-up and next transfusion

3.0 References

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