1.0 Introduction

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.

2.0 Background

Cerebrovascular accident (CVA) occurs in 5–10% of people with Sickle Cell Anaemia. The risk of CVA is highest in such children between 1 and 9 years of age. Infarctive strokes are more common in children, whereas hemorrhagic strokes occur more frequently in adults (ages 20–29). Marked stenosis or obstruction of the anterior or middle cerebral arteries are the most common abnormalities found in children.

Thrombosis and intimal hyperplasia, the precursors of infarctive 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 tears in over-dilated vessels. The risk of infarctive 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%).

3.0 Clinical Features

- **Ischemic stroke** typically presents acutely with signs and symptoms of hemiparesis or hemi-anesthesia, 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 generalized phenomena such as coma, headaches, and seizures.

- **Transient ischemic attacks (TIA)** are defined by neurological signs that resolve within 24–48 hours; they often occur before an infarctive stroke, but may go unnoticed in young children.

**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.
4.0 Recommendations for Emergency Department Treatment

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

**Child with Sickle Cell Disease presents with suspected Stroke (CVA) in ED**

- Provide immediate assessment and management including:
  - Stabilize vital signs
  - Provide life support if indicated
  - Administer oxygen to maintain \( O_2 \) saturation > 95%
  - Treat seizures and increased intracranial pressure if indicated

**Initiate stroke protocol in Hyperacute Arterial Ischemic Stroke Pathway**

- Note: TPA is contraindicated in patients with stroke secondary to sickle cell

**Gather history and complete physical exam**

- Consult: Haematology fellow 
- Stroke or Neurology fellow/staff on call
*Haematology fellow should see all sickle cell patients with stroke and discuss with staff

- Keep NPO and establish IV fluid maintenance
  - *add IV fluids with dextrose to avoid hypoglycemia as per NPO order (Serum electrolytes should be ordered prior to IV fluid administration as per 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
  - Note: CT scan during ED visit may appear normal; CT scan conducted 2-7 days post CVA usually shows areas of infarction

- 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).
  - Note: if the child is febrile, refer to Fever Guidelines for Management in Children with Sickle Cell Disease

- If moderate to severe pain, refer to the Acute Painful Episodes Vaso-occlusive Crisis: Guidelines for Management in Children with Sickle Cell Disease, and accompanying order set.

- Begin the exchange transfusion preparation
  - Exchange transfusion will take place in CCU

**End of ED Management Recommendations**

- CCU and Inpatient Recommendations on next page

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5.0 Recommendations for In-patient Management: Critical Care Unit and Ward

Admit to CCU 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 CVA, and/or clear history/physical indicating CVA: perform double-volume RBC exchange transfusion to a haemoglobin of 100g/L, and HbS level of <30% of total Hgb (see Exchange Transfusion Protocol).

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 to rule out Right to Left shunt prior to discharge

End of CCU Management

Paediatric 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

Discharge when all criteria below are met:

- The patient has been clinically and neurologically stable for at least 48 hours post transfusion(s);
- The child has been afebrile for at least 24 hours;
- The child is taking fluids and medications orally;
- Haematology, stroke service and physical therapy follow-up has been organized; and
- A plan for chronic transfusion program is in place.
Stroke: Guidelines for In-patient Management in Children with Sickle Cell Disease

6.0 References


7.0 Guideline Group and Reviewers

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Appendix A: Grades of Recommendation

- A: Recommendation supported by at least one randomized controlled trial, systematic review or meta-analysis
- B: Recommendation supported by at least one cohort comparison, case study or other experimental study
- C: Recommendation supported by expert opinion or experience of a consensus panel

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# Stroke: Guidelines for In-patient Management in Children with Sickle Cell Disease

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## Attachments:

- [sickle cell_ED_March 23.pdf](sickle_cell_ED_March_23.pdf)
- [Exchange Transfusion Calculation.pdf](Exchange_Transfusion_Calculation.pdf)
- [sickle cell_inpatient_final_April 17.pdf](sickle_cell_inpatient_final_April_17.pdf)
- [Stroke Protocol.pdf](Stroke_Protocol.pdf)
- [Revision History.docx](Revision_History.docx)