

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

Version: 3

This is a CONTROLLED document for internal use only, valid only if accessed from the Policies and Procedures site.

### 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.

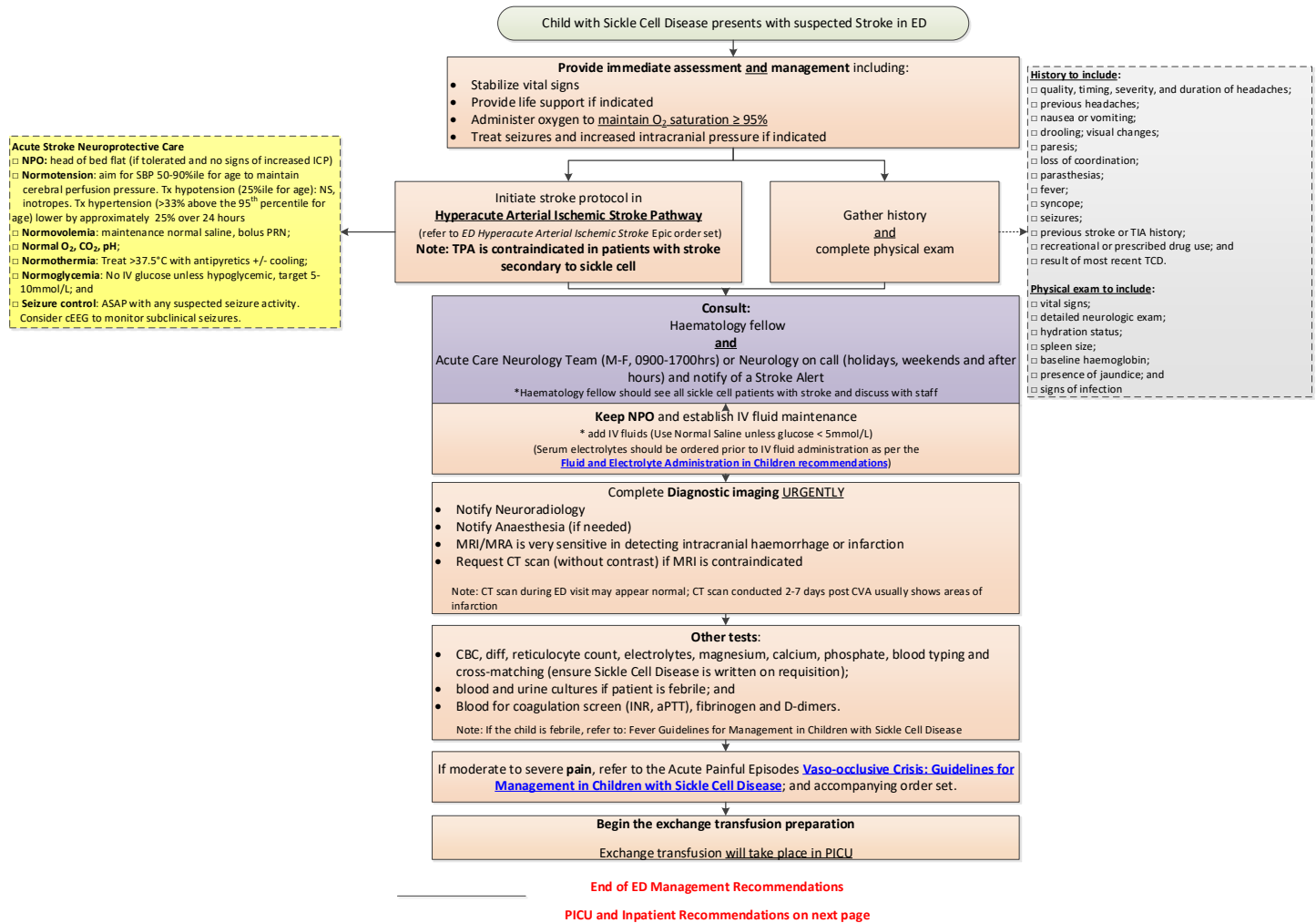
### Clinical Features

- **Arterial Ischemic stroke** typically presents acutely with signs and symptoms of hemiparesis or 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 [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

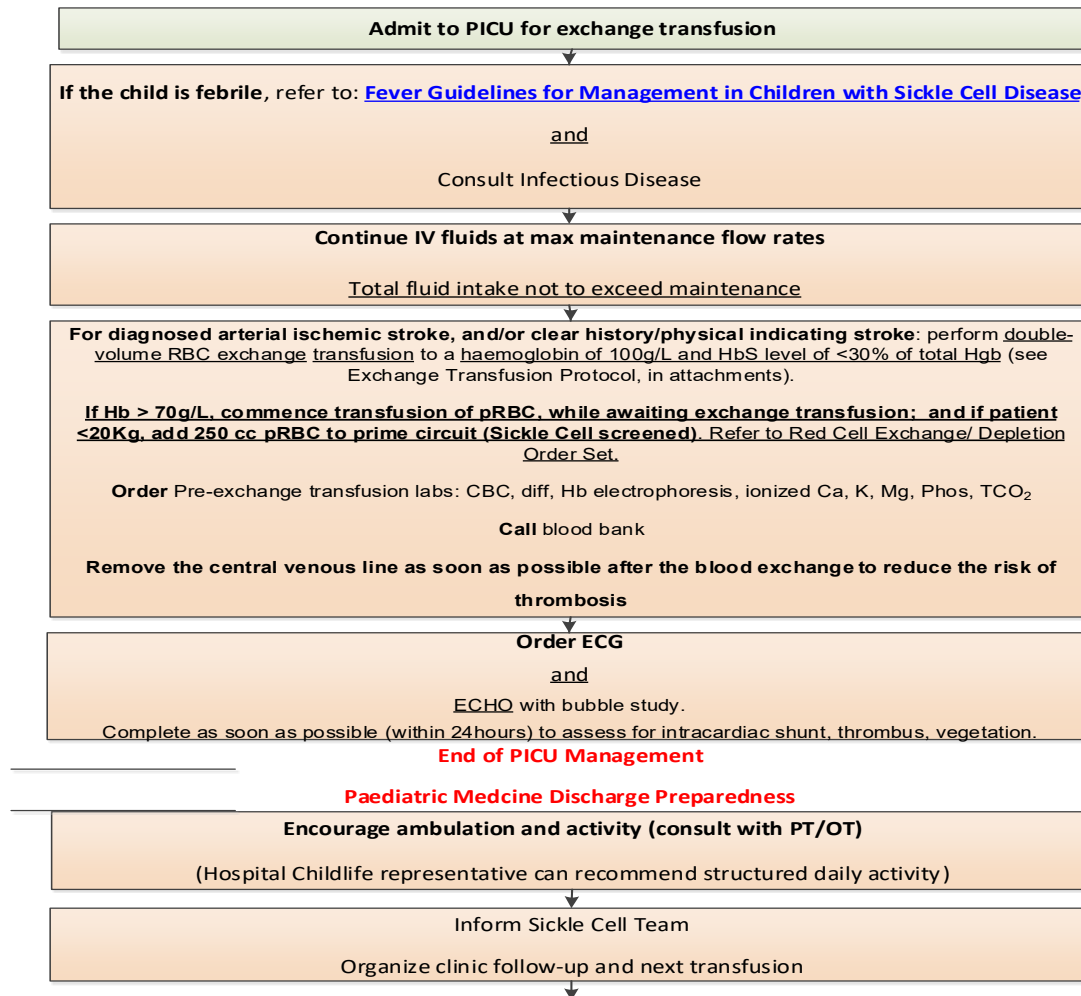


## Printable version

## 5.0 Recommendations for In-patient Management: PICU and Ward

©The Hospital for Sick Children ("SickKids"). All Rights Reserved. This document was developed solely for use at SickKids. SickKids accepts no responsibility for use of this material by any person or organization not associated with SickKids. A printed copy of this document may not reflect the current, electronic version on the SickKids Intranet. Use of this document in any setting must be subject to the professional judgment of the user. No part of the document should be used for publication without prior written consent of SickKids.





## [Printable version](#)

## References

1. Adams R, McKie V, Nichols F, Carl E, Zhang D, McKie K, Figueroa R, Litaker M, Thompson W, Hess D. The use of transcranial ultrasonography to predict stroke in sickle cell disease. *N Engl J Med*. 1992;326(9):605–10.
2. Balkaran B, Char G, Morris JS, Thomas PW, Sergeant BE, Sergeant GR. Stroke in a cohort of patients with homozygous sickle cell disease. *J Pediatr*. 1992;120(3):360–66.
3. Ohene-Frempong K, Weiner S, Sleeper L, Miller S, Embury S, Moehr J, Wethers D, Pegelow CH, Gill F. Cerebrovascular accidents in sickle cell disease: rates and risk factors. *Blood*. 1998;91(1):288–94.
4. Pegelow CH, Adams RJ, McKie V, Abboud M, Berman B, Miller ST, Olivieri NF, Vichinsky E, Wang W, Brambilla D. Risk of recurrent stroke in patients with sickle cell disease treated with erythrocyte transfusions. *J Pediatr*. 1995;126(6):896–99.
5. Reid CD, Charache S, Lubin B (eds). *Management and Therapy of Sickle Cell Disease, 3rd edition*. National Institutes of Health Publication No. 95-2117, Bethesda, Maryland, 1995.

©The Hospital for Sick Children ("SickKids"). All Rights Reserved. This document was developed solely for use at SickKids. SickKids accepts no responsibility for use of this material by any person or organization not associated with SickKids. A printed copy of this document may not reflect the current, electronic version on the SickKids Intranet. Use of this document in any setting must be subject to the professional judgment of the user. No part of the document should be used for publication without prior written consent of SickKids.

6. Russell M, Goldberg H, Hodson A, Kim H, Halus J, Reivich M, Schwartz E. Effect of transfusion therapy on arteriographic abnormalities and on recurrence of stroke in sickle cell disease. *Blood*. 1984;63(1):162–69.

**Reviewers (listed alphabetically):**

1. Carolyn Beck, MD, Staff Paediatrician, Division of Paediatric Medicine
2. Ishvinder Bhathal, NP, Neurology
3. Melina Cheong, RN, Nurse Practitioner, Division of Haematology/Oncology
4. Joy Mburu MD, Haematology/Oncology Fellow
5. Mahendra Moharir, MD, Staff Neurologist
6. Olivia Ostrow, MD, Staff Paediatrician, Division of Paediatric Emergency Medicine
7. Marcia Palmer, RN, Division of Haematology/Oncology
8. Suzan Williams MD, Physician , Division of Haematology/Oncology

**Attachments:**

[Exchange Transfusion Calculation.pdf](#)

[Revision History.docx](#)

[sickle cell ED Pathway 2021 Final.pdf](#)

[sickle cell inpatient Pathway 2021 FINAL.pdf](#)

[Stroke Protocol 2021.pdf](#)