Scope: Hospital-wide Patient Care
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Stroke: Guidelines for In-patient Management in Children with Sickle Cell Disease

Version: 4

1.0 Introduction

Stroke and the cognitive sequelae represent a source of significant morbidity in sickle cell disease. By age 20 years, strokes occur in 11% of patients with HbSS, and 4% of patients with Hb SC disease. The risk of stroke is highest 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 strokes 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.

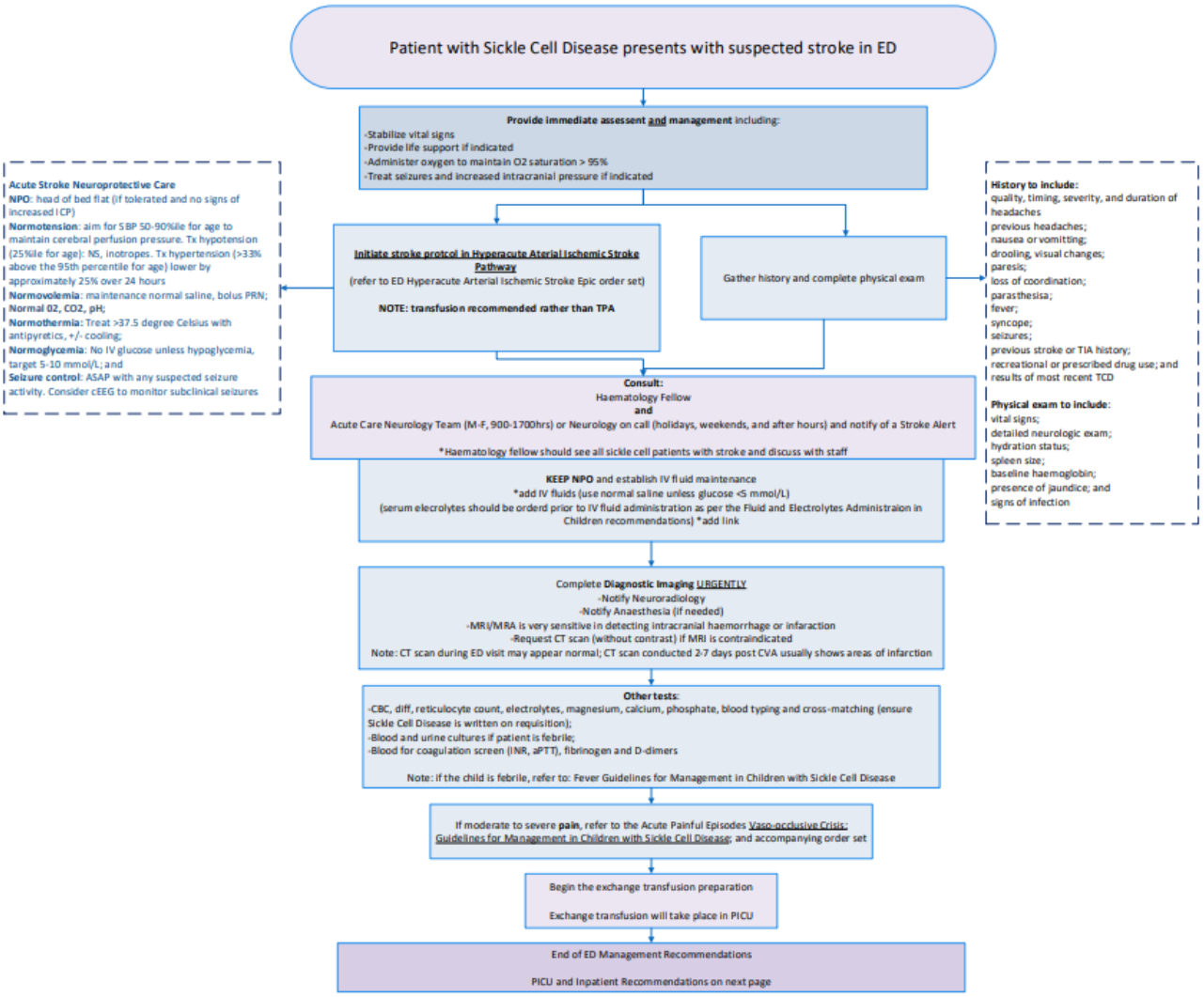
2.0 Clinical Features

- **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.

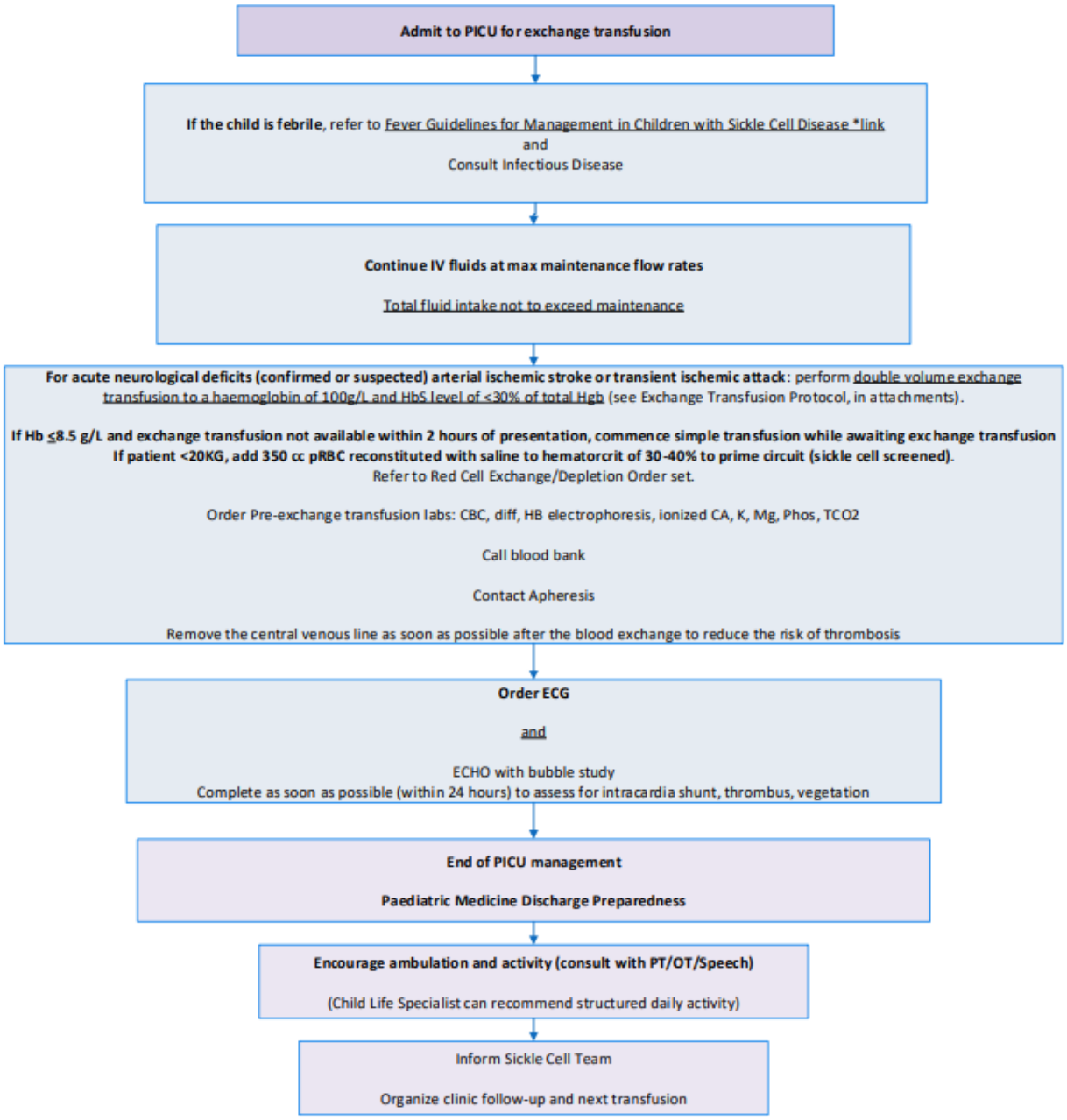
- **Transient ischemic attacks (TIA)** are defined by neurological signs that resolve within 24–48 hours; they are often a precursor to arterial ischemic stroke and should be treated as an emergency.

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.

3.0 Recommendations for Emergency Department Treatment



4.0 Recommendations for In-patient Management: PICU and Ward



Link to Pathway

5.0 References

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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. Mahendra Moharir, MD, Staff Neurologist
5. Olivia Ostrow, MD, Staff Paediatrician, Division of Paediatric Emergency Medicine
6. Marcia Palmer, RN, Division of Haematology/Oncology
7. Suzan Williams MD, Physician , Division of Haematology/Oncology

Attachments:

Exchange Transfusion Calculation.pdf

Revision History.docx