Sickle Cell Disease: Stroke ED/Inpatient

Associated Power Plans: EDP Sickle Cell Stroke (Suspected); Sickle Cell Stroke (Suspected)

Children's Mercy KANSAS CITY | Evidence Based Practice

Inclusion criteria:
- Child diagnosed with sickle cell disease (SCD) presenting with signs/symptoms of a suspected stroke

Note. If child is known to CMKC, review the Critical Information note and type of SCD (HbSS and HbSβ⁰ thalassemia have a higher risk of stroke than HbSC or HbSβ+)

History
- Stroke, transient ischemic attack
- Headaches
- Nausea or vomiting
- Visual changes
- Weakness
- Loss of coordination
- Numbness and tingling
- Fever
- Syncope
- Seizures
- Recreational or prescribed drug use

Physical Exam
- Baseline mental status with detailed neurologic exam
- Hydration status
- Signs of infection

Differential Diagnosis
- Complicated migraine
- Posterior reversible encephalopathy syndrome
- Seizure
- Cerebral venous sinus thrombosis, refer to *Cerebral Venous Sinus Thrombosis Clinical Pathway*
- Hemorrhagic stroke
- Meningitis
- Sepsis, refer to *Sepsis Clinical Pathway*
- Vaso-occlusive crisis, refer to *Sickle Cell Disease: Management of Acute Pain Clinical Pathway*

This list is not all inclusive of possible differential diagnoses

Acute Sickle Cell Stroke Neuroprotective Care
- Head of bed flat, if tolerated and there are no signs of increased intracranial pressure
- Avoid hypotension: Bolus as needed with NS 10 - 20 mL/kg
- Normovolemia: NS at maintenance ***-or-*** D5NS if glucose < 100
- Saturations > 96%
- Normothermia: Treat temperature > 38°C with antipyretics, with or without cooling blanket
- Seizure control:
  - As soon as able with any suspected seizure activity
  - Consider continuous EEG to monitor subclinical seizures (*consult Neurology as soon as able for seizure prophylaxis recommendations*)

Child with sickle cell presents with signs/symptoms of a suspected stroke (ED/Inpatient)
Activation of Stroke Alert is not indicated

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History and Physical
- Obtain history and complete physical exam
- Consider differential diagnosis

Immediate Stabilization Management
(Use EDP Sickle Cell Stroke [Suspected]or Sickle Cell Stroke [Suspected] Powerplan)

If patient presents to CMK ED, plan to immediately transfer to Adele Hall for imaging/transfusion as soon as possible after initial stabilization, IV placement, and labs

- **Provide supplemental oxygen:** Maintain oxygen saturation levels above 96%
- **Monitor vital signs:** Check vital signs, including blood pressure, every 15 minutes for the first hour
- **Provide neuroprotective care**
- **NPO status and IV access**: Keep the child NPO (nothing by mouth) and establish IV access
- **Pain management:** Ensure adequate pain control
- **Prepare for PICU admission:** Notify the PICU early for admission and potential RBC exchange
- **Consultations**: (1) Contact PICU and consult Hematology/Oncology early to help guide evaluation process and treatment; (2) Involve Apheresis Team; and (3) Consult Neurology if seizures present or if presentation unclear or not entirely consistent with stroke
- **Order MRI:** Use Stroke protocol imaging embedded in order set
- **Place Blood Bank and laboratory tests orders:**
  - **Blood Bank**: Type and cross-match for sickle-negative PRBCs (*order 1 unit for child < 30 kg and 1 - 2 units for child > 30 kg*), hold blood as per blood bank for extended phenotype
  - **Laboratory tests**: CBC with differential, reticulocyte count, hemoglobin S, BMP, Mg, iCa, Phos, PT, INR, aPTT, LFT, fibrinogen, D-Dimer, urine hCG (*for female > 10 years of age*) or serum beta hCG, POC glucose
- **Fever management**: If child has a fever,
  - Obtain blood culture then treat with acetaminophen and antibiotics
    - Consider administering ceftriaxone +/- azithromycin, if concerns for acute chest

Do not delay treatment while waiting for imaging, notification from consulting providers, or while placing orders to obtain labs or get IV access - *quickly transferring to the PICU to initiate definitive treatment is encouraged, particularly if high suspicion for sickle cell stroke*

Imaging and Transfusion
- If Hgb < 8.5 g/dL, initiate transfusion of 15 mL/kg PRBCs over 2 hours
- Obtain MRI if it can be done safely ***-and-*** within 60 minutes
  - If MRI is **NOT** available within 60 minutes, obtain non-contrast head CT STAT
- If uncertain of diagnosis, request stat MRI while waiting for requested Hgb SS fraction
  - Full MRI is preferred given the broader differential
  - Encourage discussion between attending (PICU) and attending radiologist

Transfusion should not delay imaging

Does imaging identify hemorrhage, ischemic stroke, or inconclusive findings?

- **Hemorrhage** → • Consult Neurosurgery • Plan for PICU admission
- **Ischemic Stroke** → Admit to PICU and prepare for exchange transfusion once in PICU
- **Inconclusive** → Admit to PICU ***-or-*** Hematology service if child is stable