Sickle Cell Disease: Stroke
PICU/Inpatient
Associated Power Plans: Sickle Cell Stroke (Suspected)

Children's Mercy KANSAS CITY | Evidence Based Practice

[Neurocritical Care: Sickle Cell Stroke Summary]

QR code for mobile view

Child admitted to PICU for exchange transfusion due to sickle cell stroke

Treatment
- Use Sickle Cell Stroke (Suspected) admission order set
- Continue IV fluids at 1x maintenance flow rates (*total fluid intake should **NOT** exceed maintenance*)
- Provide adequate pain control
- Place pheresis catheter
- Ensure Hematology/Oncology, Neurology, and Apheresis Team consults in place

Avoid hyperosmolar therapies which may worsen sickling

Prior to Exchange
- Provide neuroprotective care
- Ensure pre-exchange transfusion labs were obtained prior to exchange
- Call Blood Bank to request cross-matching multiple units in preparation for exchange transfusion
- Call lab to confirm receipt of Hgb S (*do not wait for Hgb S level to initiate apheresis*)

For Imaging-Confirmed Sickle Cell Stroke or Suspected Based on History and Physical
- If Hgb < 5, advise simple transfusion only
- If Hgb 5 - 8.5, perform RBC exchange transfusion to a Hgb of 9 - 12 and Hb S goal of 15 - 20% of total Hgb. Perform simple transfusion if delay in exchange is expected
- If Hgb > 8.5, perform RBC exchange transfusion to a Hgb of 9 - 12 and Hb S goal of 15 - 20% of total Hgb

Upon Completion of Exchange Transfusion
- Obtain BMP, iCa, Mg, Phos, Hgb S (*batch pre- and post- apheresis Hgb S labs*)
- If appropriate (Hgb S < 30%), consider removal of the central venous catheter to reduce the risk of thrombosis

Additional ICU/Inpatient Management
- Encourage ambulation and activity
- Consult Rehab Medicine, Psychology, Speech Therapy, Physical Therapy, Occupational Therapy
- Consult Neurosurgery if child has evidence of Moyamoya Syndrome and/or concerns with steno occlusive disease on initial MRI/MRA

Prior to Discharge
Inform Sickle Cell Team in order to organize clinic follow-up and schedule next transfusion

Discharge
- Validate follow-up arrangements have been made with Sickle Cell Team, Neurology, Physical Therapy, Rehab Medicine, and Apheresis if ongoing automated exchanges are indicated
- Validate follow-up arrangements have been made with Neurosurgery (*if patient has evidence of Moyamoya*)

Pheresis Catheter Recommendations
- May refer to Children's Mercy *Partial Manual Packed Red Blood Cell Exchange* policy for venous access options and decision-making
- **If port present, contact Apheresis Team regarding its use for exchange**
- **Femoral line suggestions**:
  - < 15 kg: 8 Fr
  - 15 - 30 kg: 10 Fr
  - > 30 kg: 12 Fr
- **Internal jugular line suggestions:**
  - < 20 kg: 8 Fr
  - 20 - 50 kg: 10 Fr
  - > 50 kg: 12 Fr

Notes.
- Children < 5 kg require manual red cell exchange. Call Hematology for recommendations
- Children > 15 kg are eligible for automated exchange
- Children 5 - 15 kg will require additional blood to prime apheresis machine and require further discussion with Apheresis Team

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*)

Discharge Criteria
- Clinically and neurologically stable ≥ 24 - 36 hours post transfusion(s)
- Afebrile for at least 24 hours
- Able to take fluids and medications orally