Sickle Cell Disease
Clinical Pathway
Johns Hopkins All Children’s Hospital

Sickle Cell Disease Clinical Pathway

Table of Contents

1. Rationale
2. Pain Crisis
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
   d. Outcome Measures
3. Fever
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
   d. Outcome Measures
4. Acute Chest Syndrome
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
5. Splenic Sequestration
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
6. Stroke
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
7. Priapism
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
8. Cholelithiasis/Cholecystitis
   a. Algorithmic Pathway
   b. Laboratory/Imaging Studies
   c. Management
9. Documentation Reminders
10. References
Rationale

This clinical pathway was developed by a consensus group of JHACH Pediatric Emergency Medicine Physicians, Advanced Practice Providers, Hematologists to standardize the management of children evaluated for sickle cell disease and subsequent comorbidities in the JHACH pediatric emergency department. It addresses the following clinical questions or problems:

1. How to evaluate and manage a sickle cell disease patient with pain crisis, and when to admit?
2. How to evaluate and manage a sickle cell disease patient with a fever, and when to admit?
3. How to evaluate and manage a sickle cell disease patient with suspected Acute Chest Syndrome?
4. How to evaluate and manage a sickle cell disease patient with suspected splenic sequestration?
5. How to evaluate and manage a sickle cell disease patient with suspected CVA?
6. How to evaluate and manage a sickle cell disease patient with priapism?
7. How to evaluate and manage a sickle cell disease patient with cholelithiasis/cholecystitis?
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Sickle Cell Disease Pain Crisis Clinical Pathway

1st 30 minutes:
- Pain >/= 7, or severe
  - Start IV, CBC, Retic Count, CMP, UHcg (F >11 years) and check labs/imaging as indicated by history and physical exam
  - Give IV pain medications (narcotic and Ketorolac) and IVF
  - Place patient on continuous pulse oximetry

IV medications:
- Morphine 0.1-0.15mg/kg/dose (max 10mg)
  or
- Hydromorphone 0.02-0.05mg/kg/dose (max 1.5mg)
  and
  *Ketorolac 0.5mg/kg/dose (max 30mg)*

If UNABLE to obtain IV:
- Fentanyl IN 1.5mcg/kg (max 100mcg)
- Morphine SC 0.1-0.15mg/kg (max 10mg)
- Oxycodeone PO 0.2mg/kg (max 10mg)
  Also consider oral morphine or oral Dilaudid

If no concern for acute chest:
20cc/kg NS bolus over 60 minutes, IVF 1.5x maintenance

31-60 minutes:
- Reassess pain

Pain Improved:
- If patient comfortable managing pain at home, discuss with Hematology team
- Discharge home with home pain plan/regimen

61-90 minutes:
- Reassess pain

Pain: Severe or 7 or above
- Give 2nd dose of narcotic pain medication, nursing standing order if patient is alert, responsive

Pain Improved:
- If patient comfortable managing pain at home, discuss with Hematology team
- Discharge home with home pain plan/regimen

Pain: Severe or 7 or above:
- Give 3rd dose of narcotic pain medication
- Proceed with admission process
- Consult hematology team, admit under hematology attending to inpatient status

Consider Dx codes:
- Hb-SS with Crisis
- Thalassemia with Crisis
* Please add pain location as indicated

*Contraindications to Ketorolac:
- Pregnancy
- Renal impairment
- Last dose of ketorolac within 5 days
- Last dose ibuprofen within 6 hours

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<table>
<thead>
<tr>
<th><strong>Sickle Cell Disease (SCD) Pain Crisis Minutes 0-30</strong></th>
<th><strong>Sickle Cell Pain Crisis Minutes 31-60</strong></th>
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</table>
| - Do a thorough physical exam and gather recent pain history and home meds used (i.e., has patient tried to adequately treat pain with oral regimen)  
- Evaluate pain with pain scale  
- If pain level 7 or above, give a dose of pain medication, preferably IV  
- Place patient on continuous pulse oximetry  
- Pain medication dosing  
  - Morphine 0.1-0.15 mg/kg/dose (max 10mg)  
  - Hydromorphone (Dilaudid) 0.02-0.05 mg/kg/dose (max 1.5 mg)  
  - If unable to get IV access, consider IN fentanyl 1.5mcg/kg (max 100mcg), SC morphine 0.1-0.15mg/kg (max 10mg) or PO oxycodone 0.2mg/kg (max 10mg); also consider PO morphine or PO Dilaudid  
  - In addition to IV narcotic pain medicine, give Ketorolac 0.5mg/kg/dose (max 30mg) IV if patient is not pregnant, does not have renal impairment and has not had Ketorolac in the last 5 days; confirm with patient/family when last dose of ibuprofen was given (wait 6 hours between doses).  
  - Check labs: CBC, retic, CMP, Urine HCG (females >11 years of age)  
  - Check other labs as indicated by history and physical exam  
  - If there is NO suspicion or evidence of acute chest syndrome, give a 20ml/kg NS bolus over 60 minutes and then start IVF at 1½ times maintenance. If acute chest syndrome suspected or evident, start IVF at maintenance rate and avoid bolus/excess fluid load unless clinically indicated  
  - Warm blankets and warming packs as needed  
  - Obtain a two-view CXR if patients has chest or upper back pain. | - Reassess pain  
- If pain has improved and patient is comfortable managing pain at home, then discuss with Hematology Team and discharge home with home pain plan/regimen  
- If pain level 7 or above, give 2nd dose of narcotic pain medication; nursing standing order to give 2nd IV narcotic pain medication if patient is alert and responsive | - Reassess pain  
- If pain has improved and patient is comfortable managing pain at home, then discuss with Hematology Team and discharge home with home pain plan/regimen  
- If pain level is 7 or above, give a 3rd dose of narcotic pain medication and proceed with admission process  
- Call Hematology service to discuss with attending  
- If patient is admitted, admit under Hematology attending with inpatient status if patient is safe for the floor |
Laboratory/imaging studies: A patient undergoing evaluation and management for a vaso-occlusive pain crisis due to sickle cell disease should have a CBC, reticulocyte count, CMP drawn (Evidence Low, consensus national panel of experts along with local expert recommendation). For females >11 years of age, a urine Hcg should be checked (Evidence Low, local expert recommendation), especially if Ketorolac may be considered for pain management as it is contraindicated in pregnancy. Obtain a 2-view chest x-ray if patient has chest or upper back pain to evaluate for cause of pain or for acute chest syndrome, especially if it is not their usual pain crisis location (Evidence Low, local expert recommendation).

Pain management: Vaso-occlusive pain crisis from sickle cell disease can be difficult to evaluate and manage. Emergency department clinical pathways have shown to improve time to pain assessments, time to pain medication administration, pain management and discharge rates (Evidence High, multiple observational trials). It is imperative to administer pain medication quickly, within 30 minutes of triage, as decreased time to pain management has been shown to improve patient outcome (Evidence Low, Mathias MD, McCavitt TL Timing of opioid administration as a quality indicator for pain crises in sickle cell disease, consensus national panel of experts). As long as it is not contra-indicated, IV Ketorolac should be given for pain control along with IV narcotics for severe acute pain crisis management (Evidence low, Beiter et al and local expert recommendation). IVF bolus and continuous IVF should be administered if the patient appears dehydrated or has poor PO intake (Evidence Low, local expert recommendation). Pain levels should be assessed every 30 minutes, with more pain medication administered as needed for severe pain (Evidence Low, consensus national panel of experts).

Admission: Patients with sickle cell disease who have pain crisis which they are unable to manage at home or whose pain levels are still moderate to severe despite adequate pain medications in the emergency department should be admitted to the hematology service under inpatient status for further pain control. Diagnostic codes to consider include Hb-SS with Crisis, Thalassemia with Crisis. Please include the body part or location of the pain when possible.

Outcome measures: Key measures include: Time to 1st narcotic pain medication administration, time to 2nd narcotic pain medication administration, % patients with CXR, length of stay in emergency department for admitted patients, length of stay for discharged patients, time to decision to admit (consult hematology), admission rate.
Sickle Cell Disease with Fever Algorithmic Pathway

**Sickle Cell with fever >100.4**

1st 60 minutes:
- SpO2 monitor, Start IV, CBC, CMP, retic, blood cultures (peripheral plus central line blood cultures from all lumens if present), UHcG for females >11 yrs
- Source of fever/tests to consider: Strep, UA, RVP, Flu, CXR, osteo

**Concern for Acute Chest Syndrome?**
- Chest/back pain, cough, tachypnea, dyspnea, hypoxia, increased WOB, abnormal lung findings

**Acute Chest Syndrome**
- IV or IM Ceftriaxone 50mg/kg dose within 60 minutes
- Azithromycin 10mg/kg, (max 500mg)
- Limit IVF, consider only 3/4-MIVF; avoid IVF bolus unless indicated then consider 10cc/kg bolus
- See ACS Guideline and Pathway

**No signs of ACS:**
- IV or IM Ceftriaxone 50mg/kg (max 2g), within 60 minutes
- If unstable add Vancomycin 15mg/kg (Max 1g)
- If Cephalosporin allergy, use Ampicillin 50mg/kg/dose (max 2000mg)
- If Cephalosporin and PCN allergy, give Levaquin 8-10mg/kg/dose IV for patients 6 months to <= 5 years, and Levaquin 10mg/kg/dose for patient > 5 years
- IVF bolus 20cc/kg over 60 minutes

**Admission Criteria:**
- Age <1 year
- Fever >39.5C AND Ill appearing,
- Poor compliance with medications
- Inability to follow up
- Prior bacteremia/sepsis
  - WBC >30k
  - Hgb <5g/dL
  - Infiltrate on CXR
  - Need for O2

Admit to Hematology
*Inpatient status to hematology/oncology floor*
Sickle Cell Disease with fever

- Physical exam and history (duration and degree of fever, antipyretic use, associated infectious symptoms, penicillin compliance if applicable, h/o splenectomy, etc.), O2 sat
- Check labs: Peripheral blood culture and central line culture from all lumens if the patient has a central line, CBC, retic, CMP, Urine HCG (as indicated)
- Obtain two-view CXR if respiratory symptoms, history of Acute Chest Syndrome or leukocytosis >18,500
- Evaluate possible source of fever and test accordingly (strep pharyngitis, Ua/UCx, influenza, osteomyelitis, respiratory viral panel PCR, etc.)
- Start empiric antibiotics ASAP (within 60 minutes): IV/IM Ceftriaxone 50mg/kg (max 2000mg); if Cephalosporin allergy, use Ampicillin 50mg/kg/dose IV (max 2000mg) and if the patient gets discharged home, then prescribe high dose amoxicillin 90mg/kg/day divided TID for 24 hours; if Cephalosporin and PCN allergy, use Levaquin 8-10mg/kg/dose IV for patients 6 months to <5 years with one IV dose in the ED and one PO dose to be given at home 12 hours later if the patient is discharged, and for patients >=5 years give Levaquin 10mg/kg/dose q24 hrs IV; If the patient is unstable, add Vancomycin 15mg/kg (max 1gm)
- DO NOT wait for lab or radiology results to administer antibiotics
- IV fluid bolus 20 cc/kg or more depending on fluid status, BP; if acute chest syndrome suspected or evident, start 3/4-MIVF and avoid bolus/excess fluid load unless clinically indicated
- Criteria for definite admission with fever: Age < 1 year, fever >39.5 AND ill-appearing, poor compliance with clinic follow up/immunizations/penicillin (younger child), prior bacteremia/sepsis, WBC > 30k, Hgb <5 g/dL, infiltrate on CXR or O2 requirement
- Discuss with Hematology re: admission. If admitted, admit under hematology service, inpatient status if patient is safe for the floor

Lab/imaging: A patient undergoing evaluation and management for sickle cell disease with fever should have a blood culture drawn. If the patient has no central line, then a peripheral blood culture must be obtained. If the patient has a central line, a peripheral blood culture is still recommended, along with a blood culture from every lumen of the central line (Evidence low, consensus national and local experts). CBC, reticulocyte count, cmp should be drawn (Evidence Low, consensus national panel of experts along with local expert recommendation). For females >11 years of age, a urine Hcg should be checked (Evidence Low, local expert recommendation), especially if Ketorolac may be considered for pain management if the patient has pain crisis also as it is contraindicated in pregnancy. Obtain a 2-view chest x-ray if the patient has respiratory symptoms, chest or back pain, history of acute chest syndrome or leukocytosis of >18,500 (Evidence Low, local expert recommendation). Other studies should be obtained as warranted by history and physical exam, including urine studies, strep pharyngitis, influenza, RSV, respiratory viral panel, osteomyelitis.
**Management:** Patients with sickle cell disease and fever are at high risk for serious bacterial illness, especially by encapsulated gram-negative bacteria, i.e. streptococcus pneumoniae. Although the risk has decreased since the widespread use of vaccines, the risk for serious bacterial illness is still substantial for those patients with sickle cell disease. Antibiotics should be started within 60 minutes of patient’s arrival, with a 3rd generation cephalosporin, i.e. ceftriaxone, for the gram-negative bacteria coverage (Evidence low, consensus national and local experts). If there is a cephalosporin allergy and no documented penicillin allergy, then give Ampicillin 50mg/kg/dose (max 2000mg) IV. If the patient can be discharged home from the ED, prescribe high dose Amoxicillin 90mg/kg/day divided TID for 24 hours. If the patient has a cephalosporin allergy and a penicillin allergy, give Levaquin 8-10mg/kg/dose IV for patients 6 months <= 5 years and if the patient is discharged home, then Levaquin 8-10mg/kg/dose given PO 12 hours later. For patients >= 5 years, give Levaquin 10mg/kg/dose IV; no home dose is required (Evidence low, consensus national and local experts). Meropenem is also an option, but should be used in conjunction with hematology recommendations (Evidence low, local expert recommendation).

**Admission:** The risk of bacteremia is low in well appearing patients without other sources of infection (Evidence low, Shihabuddin BS, Scarfi CA Fever in children with sickle cell disease: are all fevers equal, Bansil NH, Kim TY, Tieu L Incidence of serious bacterial infections in febrile children with sickle cell disease, national and local expert recommendation). Criteria for admission for a patient with sickle cell with fever includes, but is not limited to: age <1 year, fever >39.5 and ill-appearing, poor compliance with clinic follow-up, poor compliance with penicillin prophylaxis, incomplete immunizations, prior bacteremia or sepsis, WBC > 30,000, Hgb < 5, infiltrates on CXR or oxygen requirements (Evidence low, national and local expert recommendation). Since many sickle cell disease patients are functionally asplenic by 2-5 years of age, consider in your medical decision making the diagnosis or possible diagnosis of Fever in asplenic Hb-SS patient. For diagnostic codes, the source of the fever is the primary diagnosis. Please include secondary diagnoses such as Hb-SS, sickle cell disease without crisis. Also consider diagnostic codes for acquired asplenia, functional asplenia, h/o asplenia.

**Outcome measures:** Should improve key outcome measures such as time to antibiotic, admission rate, length of stay in ED, percentage of patients whom a chest x-ray is obtained.
Concern for Acute Chest Syndrome?
Chest/back pain, cough, tachypnea, dyspnea, hypoxia, increased WOB, abnormal lung findings

1st 60 minutes:
- SpO2 monitor, Start IV, CBC, CMP, Retic Count, UHcg (as indicated), 2 view CXR
- Other source of fever/tests to consider: Strep, UA, RVP, Flu
- Blood Cultures (peripheral culture and all lumens of central line)
- IV or IM Ceftriaxone 50mg/kg (max 2) within 60 minutes (DO NOT WAIT FOR CXR)
- PO Azithromycin 10mg/kg (max 500mg) on day 1
  then 5mg/kg (max 250mg) Qday on days 2-5
- If Cephalosporin allergy, use Ampicillin 50mg/kg/dose (max 2000mg)
- If Cephalosporin and PCN allergy, give Levaquin 8-10mg/kg/dose IV for patients 6 months to <= 5 years, and Levaquin 10mg/kg/dose for patient > 5 years

Other Treatments:
- Albuterol PRN for wheezing
- Incentive Spirometer q1hr while awake (bubbles for young pts)
- IVF: 3/4-1x maintenance rate or less if tolerating PO. Avoid IVF bolus.
  - O2 for saturations <92%

Admission:
- Discuss with hematology
- Consider PICU for increased WOB, fatigue, increased O2 requirement, need for full exchange transfusion

Consider diagnostic codes: Hb-SS with acute chest syndrome

Inpatient Status
Acute Chest Syndrome (ACS)

- Acute chest syndrome (ACS)- suspected with fever, cough, tachypnea, chest/back pain, increased work of breathing, hypoxia, abnormal lung sounds
- Obtain two-view CXR; if patient febrile, administer IV antibiotics ASAP (obtain blood culture/labs with IV placement) and DO NOT WAIT for CXR or lab results
- Discuss with Hematology attending
- If concerned for ACS:
  - Get BCx (peripheral and all lumens of central line if present), CBC, CMP, reticulocyte count
  - Treat with IV/IM Ceftriaxone 50mg/kg (max 2000mg) and PO Azithromycin 10mg/kg (max 500mg) on day 1, then 5mg/kg (max 250mg) on days 2-5
  - If Cephalosporin allergy, use Ampicillin 50mg/kg/dose IV (max 2000mg); if Cephalosporin and PCN allergy, use Levaquin 8-10mg/kg/dose IV for patients 6 months to <5 years with one IV dose in the ED, and for patients >=5 years give Levaquin 10mg/kg/dose q24 hrs IV; If the patient is unstable, add Vancomycin 15mg/kg (max 1gm)
  - Albuterol PRN for reactive airway disease
  - Incentive spirometry—bubbles or pinwheel for younger child as often as tolerated while awake, incentive spirometer q1hr while awake for older children
  - Start IVF at ¾-maintenance rate or less if taking PO well (total fluid rate should be 3/4-1x MIVF in ACS patient)
  - Continuous pulse oximetry, give oxygen if O2 saturations <92%
  - Consider PICU placement if patient has concerning respiratory status such as increased WOB with fatigue, escalating O2 requirement, or if there is consideration for full exchange transfusion as discussed with Hematology

Labs/Imaging: Infections, including pneumonia, are a significant cause of ACS, so BCx (peripheral and all lumens of central line if present), cbc, cmp and other labs as indicated by the history and physical exam should be obtained. CXR 2-view should be obtained if there is a concern for ACS, but obtaining the CXR should not delay antibiotic administration.

Management: Patients with acute chest syndrome have a high risk or morbidity and mortality so appropriate management and early recognition is vital. Acute Chest Syndrome (ACS) should be suspected in any patient with sickle cell disease with chest or upper back pain, oxygen requirement, increased work of breathing, respiratory symptoms or previous ACS. Antibiotics should be administered as soon as possible for patients with suspected ACS. Along with Ceftriaxone or a 3rd generation cephalosporin, Azithromycin should be administered to cover Mycoplasma (Evidence Low, consensus national panel of experts). If there is a cephalosporin allergy and no documented penicillin allergy, then give Ampicillin 50mg/kg/dose (max 2000mg) IV. If the patient can be discharged home from the ED, prescribe high dose Amoxicillin 90mg/kg/day divided
TID for 24 hours. If the patient has a cephalosporin allergy and a penicillin allergy, give Levaquin 8-10mg/kg/dose IV for patients 6 months <= 5 years and if the patient is discharged home, then Levaquin 8-10mg/kg/dose given PO 12 hours later. For patients >= 5 years, give Levaquin 10mg/kg/dose IV; no home dose is required. Patients with ACS have a higher risk of pulmonary edema so IVF boluses should not be administered unless the patient’s clinical condition warrants it and the IV fluids should be restricted to a rate no greater than maintenance rate (Evidence low, local and national expert consensus). Oxygen should be administered if oxygen saturations are less than 92% (Evidence Low, consensus national panel of experts). Albuterol can be administered as needed for respiratory support to determine if it helps any possible reactive airway disease component (Evidence Low, consensus national panel of experts). Incentive spirometry or blowing bubbles or a pinwheel can help prevent symptom progressions (Evidence Low, consensus national panel of experts).

Admission: Patients with suspected ACS should be admitted under inpatient status for continued observation and further management (Evidence Low, consensus national panel of experts). Consider diagnostic code: Hb-SS with acute chest syndrome
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Splenic Sequestration Algorithmic Pathway

**Signs and Symptoms of Splenic Sequestration**

- Enlarged/tender spleen, jaundice, tachycardia, abdominal pain/distention, decreased Hgb by >\=2g/dL or more from baseline (+/- thrombocytopenia)

**Workup and Management**

- Start IV, CBC, CMP, Retic, Type & Screen, Urine HCG (as indicated)
- Pain management as per Sickle cell pain crisis pathway
- IVF@ maintenance rate
- Consider PRBC transfusion
- Consult Hematology for disposition
- Dx code: Hb-SS with splenic sequestration

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<th>Splenic Sequestration</th>
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<tr>
<td>• Suspected with enlarged/tender spleen, decrease in Hgb by 2g/dL or more from baseline, sometimes with thrombocytopenia</td>
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<tr>
<td>• Physical exam: pallor, jaundice, tachycardia, abdominal pain/distension, etc.</td>
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<tr>
<td>• Check labs: CBC, CMP, retic, type and screen, Urine HCG (as indicated)</td>
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<tr>
<td>• If concerned for splenic sequestration:</td>
</tr>
<tr>
<td>• Start IVF at maintenance rate</td>
</tr>
<tr>
<td>• Discuss with Hematology attending</td>
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<tr>
<td>• Consider PRBC transfusion</td>
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| Admission: Admit to hematology under *inpatient status*

**Labs/Imaging:** Patients with suspected splenic sequestration should have a cbc, cmp reticulocyte count drawn to evaluate for a hemoglobin drop of >\=+2g/dL form their baseline. The cbc may show thrombocytosis as well.

**Management:** Patients with sickle cell disease have an increased risk of splenic sequestration, which can manifest itself with left upper quadrant or generalized abdominal pain, enlarged spleen, anemia or jaundice. IV fluids should be run at maintenance (Evidence Low, consensus national panel of experts).
Admission: Hematology should be consulted regarding admission and further management, including pRBC transfusion. Inpatient status if admission is warranted. Diagnostic codes to consider include Hb-SS with splenic sequestration, splenic sequestration with infarct (when appropriate).
Concern for CVA:
Abnormal neurologic exam, new onset seizure activity, aphasia, altered mental status, etc.

Refer to CVA Guideline
- STAT Head CT w/o contrast and STAT Brain MRI w/wo contrast and Brain Angio MRA w/ MRV
- Check labs: CBC, retic, CMP, type and screen, hemoglobin electrophoresis (if possible)
- Discuss with Neurology re: assessment
- Discuss with Hematology re: simple or exchange transfusion
- Admit to PICU under inpatient setting
- Dx code: Acute CVA due to sickle cell disease

Lab/Imaging: Please refer to the CVA guideline. If a CVA is suspected, STAT Head CT w/o contrast and STAT Brain MRI w/wo contrast and Brain Angio MRA w/ MRV must be ordered. CBC, CMP, retic, hemoglobin electrophoresis, type and screen, along with any other studies indicated by history and physical exam (Evidence Low, consensus national panel of experts and local expert recommendations).

Management: Please refer to the CVA guideline. Sickle cell patients have a significant risk for silent and clinically apparent CVA’s due to ischemia from vaso-occlusive crisis. CVA should be suspected for any prolonged neurologic deficit (i.e. numbness, weakness, aphasia, etc.), altered mental status without other explanation (i.e. ingestion), new onset seizure activity, etc. Hematology and neurology should be consulted. Interventions, including pRBC transfusions, as indicated by hematology, neurology, etc.

Admission: Patients should be admitted to the PICU under inpatient status. Consider diagnostic code: Acute CVA due to sickle cell disease.
Priapism

- Suspected with sustained erection for > 2 hours or painful erection lasting any duration of time

- Relevant history- trauma, infections, use of medications, illicit drugs, prior occurrences
- Discuss with Hematology
- If placing an IV, obtain CBC, retic, CMP
  - If <4 hours from onset: drink fluids, take oral pain meds, and void ASAP; offer IVF and IV pain meds if not tolerating PO; can also offer PO Sudafed <12 years of age: 1 mg/kg q6hrs; ≥12 years of age 30-60mg/dose q6hrs
  - If >4 hours from onset: IV pain medication, IVF, discuss with urology for aspiration and irrigation of corpus cavernosum (under sedation) or surgical shunting

- Inpatient Status if admitted and treated

| Priapism | • Priapism- suspected with sustained erection for > 2 hours or painful erection lasting any duration of time
|          | • Obtain history- trauma, infections, medication use, illicit drugs, prior occurrences
|          | • Discuss with Hematology
|          | • If <4 hours from onset: drink fluids, take oral pain meds, and void ASAP; offer IVF and IV pain meds if not tolerating PO; can also offer PO Sudafed <12 years of age: 1 mg/kg q6hrs; ≥12 years of age 30-60mg/dose q6hrs
|          | • If >4 hours from onset: IV pain medication, IVF, discuss with urology for aspiration and irrigation of corpus cavernosum (under sedation) or surgical shunting
|          | • If placing an IV, obtain CBC, retic CMP

Labs/Imaging: CBC, CMP, Reticulocyte count if establishing IV.

Management: Patients with sickle cell disease have an increased risk of priapism, which is either a painful erection or an unwanted sustained erection lasting 2 or more hours. Untreated priapism can lead to ischemia, fibrosis, impaired sexual function and impotence. Hydration, pain control and voiding, if possible, followed by Sudafed (pseudoephedrine) are the initial steps in management for priapism < 4 hours in duration (Evidence Low, consensus national panel of experts and local expert recommendations). For priapism > 4 hours in duration, IV hydration and pain medication with a urology (Evidence Low, consensus national panel of experts and local expert recommendations). Hematology should be consulted for priapism regardless of the duration.

Admission: Meets inpatient status if treated. Consider diagnosis codes: priapism and Hb-SS
Gallbladder disease
Suspected with RUQ pain (+/- fever for cholecystitis),
intolerance of PO/vomiting (esp. fatty foods or post-prandial
pain), jaundice/icterus

- Obtain labs: CBC, retic, CMP, GGT, amylase, lipase, type
  and screen
- IVF bolus (20 cc/kg NS) and start MIVF
- IV pain medication (morphine)
- Give IV Zofran as needed for N/V
- RUQ US
- If febrile, obtain peripheral and all lumens of central line (if
  present) blood cultures and start IV Ceftriaxone 50mg/kg (max
  2gm) ASAP; Consider Zosyn if concerned for intra-abdominal
  pathology
- Discuss with Hematology re: admission (observation status
  for cholelithiasis) and surgical and/or GI consult

Cholelithiasis/Cholecystitis
- Gallbladder disease- suspected with RUQ pain (+/- fever for
  cholecystitis), intolerance of PO/vomiting (esp. fatty foods or post-
  prandial pain), jaundice/icterus
  - Check labs: CBC, retic, CMP, GGT, amylase, lipase, type and screen
  - Give IVF bolus (20 cc/kg NS) over 60 minutes and start MIVF
  - Give IV pain medication (morphine 0.1-0.15mg/kg, max 10mg)
  - Give IV Zofran as needed for N/V
  - RUQ US
  - If febrile, obtain peripheral blood culture and all lumens of central line
    blood cultures, if present, and start IV Ceftriaxone 50mg/kg (max 2gm)
    ASAP; Consider Zosyn if concerned for intra-abdominal pathology.
    - Discuss with Hematology re: admission (observation status for
      cholelithiasis) and surgical and/or GI consult

Labs/Imaging: Management for suspected gallbladder disease includes obtaining cbc, cmp, retic, amylase,
lipase, ggt and type and screen. RUQ US should be obtained to evaluate the liver, gallbladder and pancreas.
If the patient is febrile, obtain a peripheral blood culture and, if present, blood cultures from all lumens of the
central line and start IV antibiotics for concern of cholecystitis.

Management: Patients with sickle cell disease have an increased risk of gallbladder disease due to increased
hemolysis and bilirubin turnover increasing the possibility of gallstones. Gallbladder disease should be
suspected with RUQ abdominal pain, jaundice, icterus, etc. Administer IVF bolus and hydration along with IV
narcotic pain management. Consult hematology and consider GI consultation for cholecystitis and surgery consultation for cholelithiasis as a cholecystectomy is often indicated.

Admission: Observation status for cholelithiasis.

Documentation Reminders:

Please use the term “possible” when considering diagnoses in your medical decision making, such as “fever in asplenic Hb-SS patient, possible sepsis”. “Possible” is a term that can be captured as a confirmed diagnosis and later disregarded, however “suspected: or “rule out” terminology is not captured by the database those possible diagnoses would be lost or not as easily obtained.

References


Clinical Pathway Team
Sickle Cell Disease Clinical Pathway
Johns Hopkins All Children’s Hospital

Owner(s): Courtney Titus PA-C

Reviewed By:
   Hematology: Peter Shaw, MD, Jessica Wishnew, MD

Created June 2017 by: Charles Eldridge, MD, Courtney Titus PA-C, Peter Shaw, MD, Jessica Wishnew, MD

Reviewed February 2022 by Courtney Titus, PA-C

Clinical Pathway Management Team: Joseph Perno, MD; Courtney Titus, PA-C

Date Approved by JHACH Clinical Practice Council: June 2017

Date Available on Webpage: July 2017

Last Revised: February 24, 2022

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