JOHNS HOPKINS ALL CHILDREN'S HOSPITAL

Sickle Cell Disease Clinical Pathway

JOHNS HOPKINS
All Children's Hospital

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Sickle Cell Disease Clinical Pathway

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 at JHACH. 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?

Background

Sickle cell disease (SCD) is a group of inherited red blood cell disorders. Red blood cells contain hemoglobin, a protein that carries oxygen. Healthy red blood cells are round, and they move through small blood vessels to carry oxygen to all parts of the body. In someone who has SCD, the hemoglobin is abnormal, which causes the red blood cells to become hard and sticky and look like a C-shaped farm tool called a “sickle.” The sickle cells die early, which causes a constant shortage of red blood cells. Also, when they travel through small blood vessels, they get stuck and clog the blood flow. This can cause pain and other serious complications such as infection, acute chest syndrome and stroke. People with SCD may start to have signs of the disease during the first year of life, usually around 5 months of age. Symptoms and complications of SCD are different for each person and can range from mild to severe. Management of SCD is focused on preventing and treating pain episodes and other complications. Prevention strategies include lifestyle behaviors as well as medical screening and interventions to prevent SCD complications.

Sickle cell disease affects millions of people throughout the world. It is estimated that SCD affects approximately 100,000 Americans, occurring about one out of every 365 African- American births and one out of every 16,300 Hispanic-American births. Survivorship into adulthood for children with SCD has increased to more than 95% by age 18 years, and is attributed to newborn screening implementation, penicillin prophylaxis, primary stroke prevention, and disease-modifying therapies. Early diagnosis and treatment of complications of sickle cell can reduce hospitalizations and readmissions. While many complications are not preventable, the management provided in the Emergency Center and during hospitalization can improve patient outcomes and prevent additional complications from occurring.

Phenotypes of Sickle Cell Disease Included in Our Clinical Pathways:

These clinical pathways are specific for sickle cell disease. They do not include Alpha or Beta Thalassemia or Sickle Cell Trait. The most common types of sickle cell disease found in the United States are Sickle Cell Anemia (Hgb SS), Sickle Hemoglobin- C disease (HgbSC), Sickle Beta-Plus Thalassemia (S Beta-Plus), and Sickle Beta- Zero Thalassemia (S Beta- Null). These pathways should be used for any patient with sickle cell disease that presents to Johns Hopkins All Children's Hospital, regardless of disease type. If there is a question, please consult hematology.

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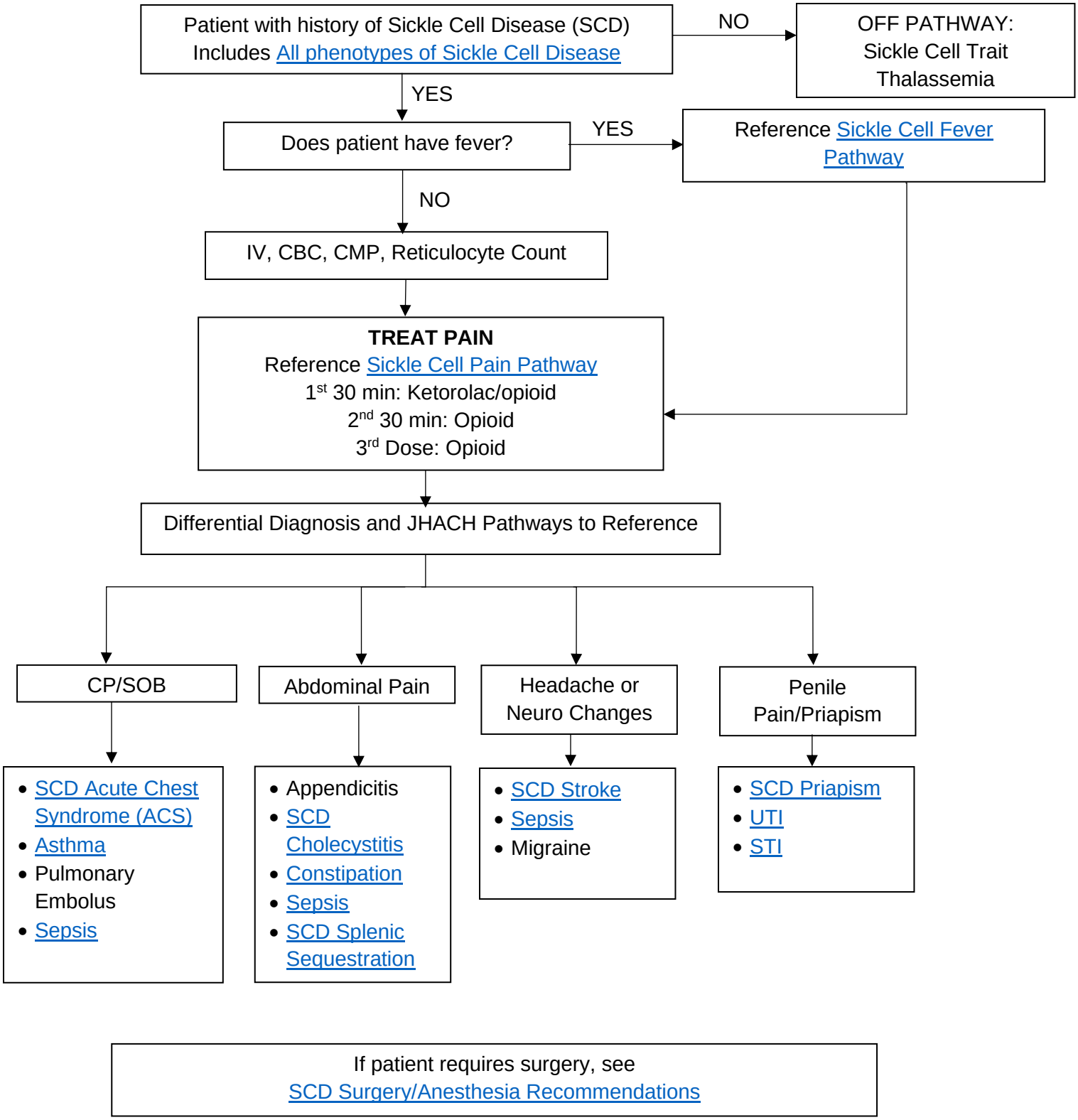
Sickle Cell Disease Initial Evaluation Clinical Pathway



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Sickle Cell Disease Pain Crisis Clinical Pathway (Emergency Center)

1st 30 minutes:
Pain ≥ 7, or unable to manage at home

- Start IV, CBC, Retic Count, CMP, UHcg (F >10 years) and check labs/imaging as indicated by history and physical exam
- Give IV pain medications (opioid and ketorolac) and IVF
- Place patient on continuous pulse oximetry
- Chest or back pain: Acute Chest Syndrome Clinical Pathway

IV medications:
Morphine 0.1-0.15mg/kg/dose (max dose: 8mg)
or
HYDROmorphone 0.02-0.05mg/kg/dose (max dose: 2 mg)
and
*Ketorolac 0.5mg/kg/dose (max dose: 30mg) *

If UNABLE to obtain IV:
Fentanyl IN 2mcg/kg (max dose: 100 mcg)
Oxycodone PO 0.2 mg/kg (max dose: 10 mg)
Also consider oral morphine or oral HYDROmorphone

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

***Contraindications to Ketorolac:**
-pregnancy
-renal impairment
-ketorolac (max 5 days out of last 30 days)
-last dose ibuprofen within 6 hours

31-60 minutes:
Reassess pain

Pain Improved:

- If patient comfortable managing pain at home, discuss with Hematology team
- Discharge home with home pain plan and 3-day oral opioid Rx

Pain: Severe or unmanageable

- Give 2nd dose of opioid pain medication
- If patient is asleep, wake and reassess pain score

61-90 minutes:
Reassess pain

Pain Improved:

- If patient comfortable managing pain at home, discuss with Hematology team
- Discharge home with home pain plan and 3-day oral opioid Rx

Pain: Severe or 7 or unmanageable:

- Give 3rd dose of opioid pain medication
- Proceed with admission process if still unmanageable
- Consult hematology team, admit under hematology attending to ***inpatient status***
- Reference Sickle Cell Disease Pain Crisis Clinical Pathway (Inpatient)

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Sickle Cell Disease Pain Crisis Clinical Pathway (Inpatient)

CONSULTS TO BE PLACED

First 24-48 hours
- Pain team
- Child life
- Music Therapy
- Social Work
- PT/ OT

48-72 hours after admission
- Academic consult
- Psychology

ACUTE CHEST SYNDROME PREVENTION

- Orders for Incentive Spirometry at least 1 time/ hour while awake
- Orders for patient to be out of bed for 1-2 hours with a goal of 4 times a day (minimum 6 hours/day)
- **Order bronchodilator Q4hr while awake with prior hx ACS or hx RAD**
- Orders for patient ambulation at least 4 times a day

NON-OPIOID PHARMACOLOGIC MANAGEMENT

- **Acetaminophen:** If < 2 yrs: 12.5 mg/kg q 6 hrs PO/PR
  If > 2 yrs. 15 mg/kg q 6 hrs PO, max dose: 1000mg, not to exceed 4g/day
  **Alternate with Ketorolac*
- **Ketorolac:** <6 months: 0.5 mg/kg q 6 hrs IV – Do not exceed 48-72 hours (max dose: 15 mg)
  ≥6 months: 0.5 mg/kg q 6 hrs IV- Give no more than 5 days in a 30-day period (max dose: 30 mg/dose)
  **Use a proton pump inhibitor or H2 antagonist to protect the GI mucosa while on NSAIDs.*

PATIENT-CONTROLLED ANALGESIA (PCA)

- Consider HYDROmorphone as first line opioid to use in PCA for sickle cell patients.
- After PCA is started, titrate the dose to the desired effect.
- Orders for PCA should be placed when it is determined that the patient will be admitted to the hematology service for VOC. PCA should be started as soon as possible after admission.
- If IV access is lost, consider PO opioids until a new IV can be placed
- If a rescue dose or clinician bolus is needed, please consult pain team or discuss with inpatient heme team attending

Has patient been on PCA in last 6 months?

YES

- Review non-opioid naive patient's individualized pain plans if available.
- Consider restarting PCA dose of previous hospitalization.

NO

- Opioid naive patients, consider following starting doses:
- HYDROmorphone: continuous rate 0.005 mg/kg/hr, bolus dose 0.003 mg/kg q 10-15 minutes
- Morphine: continuous rate 0.02 mg/kg/hr, bolus dose 0.015-0.03 mg/kg q 10-15 minutes
- Fentanyl: continuous dose 0.5 mcg/kg/hr, bolus dose 0.2-0.3 mcg/kg q 10-15 minutes

Nursing Team should reference SCD Pain Nursing Standard of Care

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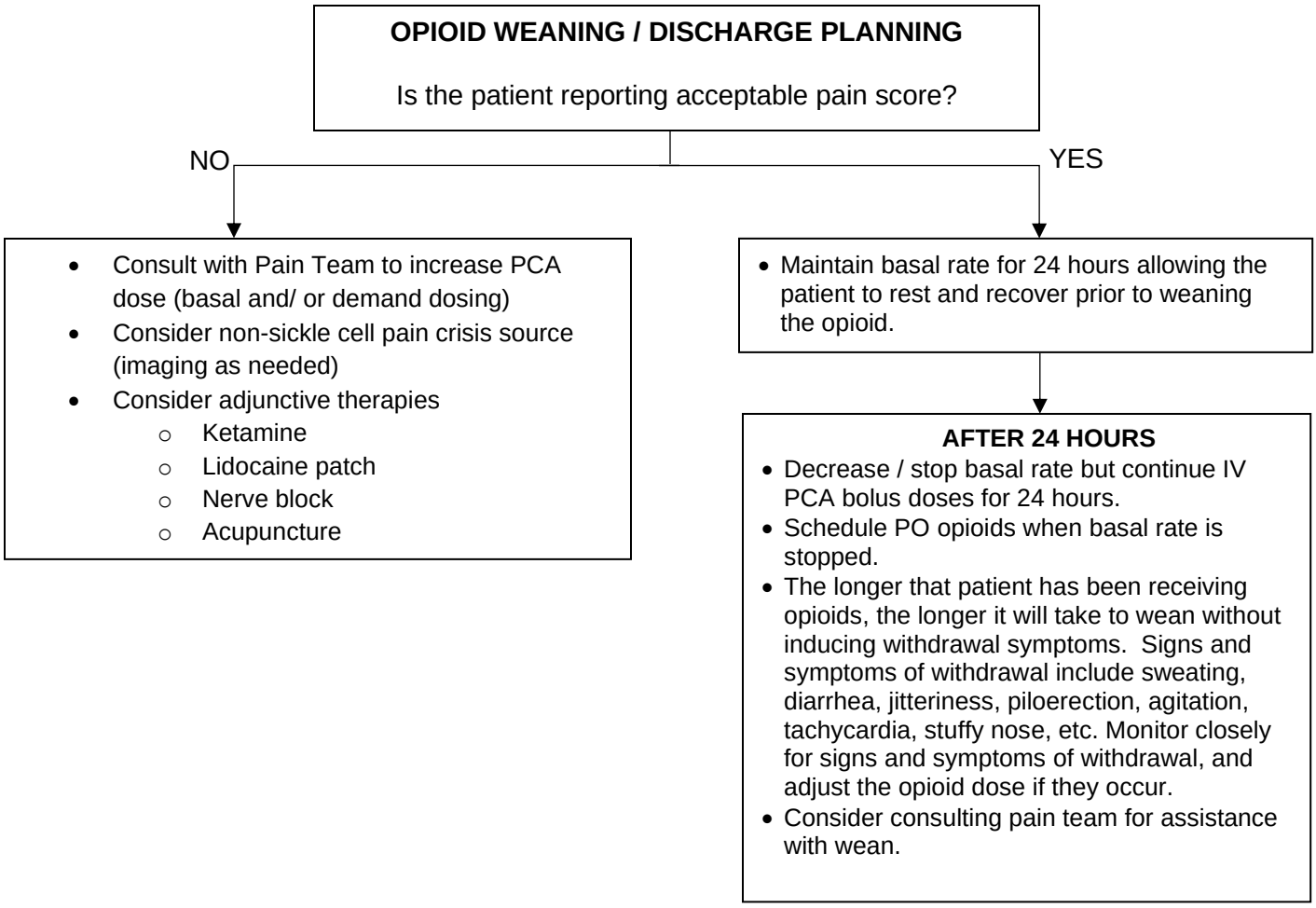
Sickle Cell Disease Pain Crisis Ongoing Management Clinical Pathway



Treatment for Common Opioid Side Effects

Constipation prophylaxis	All patients on opioid drugs for more than a day or two should receive regularly scheduled laxatives which include both a stool softener and a stimulant. Patient should have a bowel movement every 2-3 days.
Pruritus	• If pruritus develops with the use of one opioid, consider using an alternative opioid. • Butorphanol (Stadol) 0.015 mg/kg/dose IV Q4 hours (max dose: 1mg) if not effective then • Naloxone (Narcan) 0.001mg/kg IV every Q2 hours infuse over 60 min • If above medications do not control opioid induced pruritus, then start low dose naloxone IV infusion at a dose 1mcg/kg/hr and titrate up to 2 mcg/kg/hr for relief of pruritus without decreasing effectiveness of the analgesic. • **Avoid Diphenhydramine (Benadryl) (IV and oral) for opioid induced pruritus to prevent further sedation**
Nausea and vomiting	• It is first necessary to exclude a primary condition. • Ondansetron (Zofran) 0.15 mg/kg/dose (max dose: 8 mg) every 8 hours; first choice. • Promethazine (Phenergan) 0.125 mg/kg/dose (max dose: 6.25 mg) every 6-8 hours PO/IV as needed; contraindicated in children less than 2 years of age. • Lorazepam (Ativan) 0.025 mg/kg/dose (max dose: 2 mg) every 6 hours PO/IV as needed • Diphenhydramine (Benadryl) 0.5 to 1 mg/kg/dose (max dose: 50 mg) every 6-8 hours for opioid induced nausea (IV formulation highly discouraged)

Nursing Team should reference SCD Pain Nursing Standard of Care

Sickle Cell Pain Crisis

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 >10 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 opioids 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 opioid pain medication administration, time to 2nd opioid 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, return rate to EC within 72 hours or readmission within 7 days of hospital discharge.

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Sickle Cell Disease with Fever Clinical Pathway (Emergency)

Sickle Cell with fever ≥101F (38.3C)

1st 60 minutes:

- SpO2 monitor, Start IV, CBC, CMP, retic, blood cultures (peripheral plus central line blood cultures from all lumens if present),
- CXR for patients < 10 yo
- Source of fever/tests to consider: Strep, UA, RVP, Covid-19, Flu, CXR, osteo

Concern for Acute Chest Syndrome?

- Chest/back pain, cough, tachypnea, dyspnea, hypoxia,
- Increased WOB, abnormal lung findings, baseline Hgb drop > 1 g/dL

NO

YES

No signs of ACS:

- Antibiotics to be given within 60 minutes
- IVF bolus 20mL/kg over 60 minutes (max dose: 1000 mL)

Antibiotics if eligible for discharge

- cefTRIAXone 50mg/kg/dose IV ONCE (max dose: 2000 mg)
- Alternative if allergy/contraindication: levoFLOXacin
  - ≥ 6 months to <5 yrs: 10mg/kg/dose IV Once followed by 10 mg/kg/dose PO ONCE given 12 hours later (max dose: 375 mg/dose)
  - ≥ 5 yrs: 10 mg/kg/dose IV Once (max dose: 750 mg/dose)

Antibiotics if meets criteria for admission

- Ampicillin/sulbactam 75 mg/kg/dose IV ONCE (max dose: 2000 mg/dose)
- If PCN allergy: levoFLOXacin 10 mg/kg/dose IV ONCE (for patients ≥ 6 months to <5 yrs, max dose: 375 mg/dose; ≥ 5 yrs max dose: 750 mg/dose)
- Vancomycin for patients with central line, order using "JHH-ACH Pediatric Vancomycin Focused" order set

Concern for ACS

Please reference Acute Chest Pathway

Admission Criteria for Sickle Cell Fever:

- Age <1 year
- Fever >39.5C AND ill appearing
- Poor compliance with medications (penicillin)/ immunizations
- Unable or unreliable follow up
- Prior bacteremia/sepsis
- History of surgical splenectomy with unknown cause for fever
- WBC >30k or < 5k
- Hgb <5g/dL
- Infiltrate on CXR
- Need for O2
- Clinical concern

Admit to Hematology

- *Inpatient status* to hematology/oncology floor
- Any Concern for Sepsis should reference JHACH Pediatric Sepsis Pathway and admit to ICU as indicated

Nursing Team should reference SCD Fever Nursing Standard of Care

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Sickle Cell Disease with Fever Clinical Pathway (Inpatient)

Inpatient Management

- Hemodynamic monitoring, O_2 monitoring, and supportive care measures
- Continue ampicillin/sulbactam 75 mg/kg/dose IV Q6h (max dose: 2000 mg/dose) while febrile
  - If PCN allergy use levoFLOXacin*
  - If central line add vancomycin*
- Monitor blood culture for 24-48 hours**
- Re-culture with new fever if afebrile >24 hours
- Additional evaluation as needed depending on clinical symptoms
- IV plus PO to equal maintenance fluid requirement
- Incentive spirometry q1-2 hours while awake
- CXR with new fevers or if not completed in ER

Persistent fever *with abdominal/epigastric pain*, concern for **cholelithiasis, cholecystitis, hepatitis, pancreatitis**:
-Abdominal u/s, liver enzymes, amylase/lipase
- SCD Cholelithiasis/Cholecystitis Clinical Pathway

Persistent fever *with localized pain and swelling/tenderness*, concern for **osteomyelitis/septic arthritis**:
-CONSULT INFECTIOUS DISEASE
-plain film x-ray, bone scan, MRI (results can overlap with routine bone infarct)
-*Biopsy* is Gold Standard

Persistent fever with *O2 requirement, non-sternal/non-reproducible chest pain, cough, increased work of breathing*, concern for **acute chest syndrome**
- SCD Acute Chest Syndrome Clinical Pathway

Discharge Criteria

- Afebrile and blood culture negative for at least 24 hours and clinically well appearing with no other complication that requires hospital care
- Off supplemental O_2 for 24 hours, unless hx of home supplemental O_2 use
- Ensure clear method of communication in case call-back needed
- Ensure good compliance with Penicillin (if applicable)

Nursing Team should reference SCD Fever Nursing Standard of Care

*__LevoFLOXacin:__ ≥ 6 months to <5 yrs: 10mg/kg/dose IV Q12h (max dose: 375 mg/dose); ≥ 5 yrs:10 mg/kg/dose IV q24h (max dose: 750 mg/dose)
Vancomycin order using the "JHH-ACH Pediatric Vancomycin Focused" order set
**In patients with Sickle Cell Disease (SCD), time to detect bacteria in blood cultures is generally less than 24 hours

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). Obtain a 2-view chest x-ray if the patient is less than 10 years old due to increased risk of missing acute chest syndrome based on symptoms and auscultation alone. Also obtain 2-view chest x-ray if patient has respiratory symptoms, chest or back pain, concern for acute chest syndrome or hypoxia (Evidence Low, local expert recommendation). Other studies should be obtained as warranted by history and physical exam, including urine studies, strep pharyngitis, influenza, RSV, Covid-19, respiratory viral panel, osteomyelitis.

Management: Patients with sickle cell disease and fever are at high risk for serious bacterial illness, especially by encapsulated bacteria, (i.e*., Streptococcus pneumoniae, Haemophilus influenzae, Salmonella sp. and Neisseria meningitis)*. 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 ampicillin/sulbactam (or cefTRIAXone if patient meets criteria for discharge), for the gram-negative bacteria coverage (Evidence low, consensus national and local experts). If there is a penicillin allergy, then give levoFLOXacin 10mg/kg/dose IV ONCE; for patients ≥6 months to <5 years and if the patient is discharged home, then levoFLOXacin 10mg/kg/dose given PO once 12 hours later, no home dose is required for patients ≥ 5 years (Evidence low, consensus national and local experts).

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.

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Acute Chest Syndrome (ACS) Algorithmic Pathway (Emergency)

DEFINITION*

New pulmonary infiltrate on CXR involving at least one complete segment that is not consistent with appearance of atelectasis

AND

(One or more of the following)

- Chest pain
- Temp > 38.5C
- Tachypnea
- Wheezing
- Cough
- Increased WOB
- Hypoxemia relative to baseline

↓

Concern for Acute Chest Syndrome?

Chest/back pain, cough, tachypnea, dyspnea, hypoxia, increased WOB, abnormal lung sounds

↓

1st 60 minutes:

- SpO_2 monitor, Start IV, CBC, CMP, Retic Count, Type and Screen
- 2 view CXR
- Other source of fever/tests to consider: Strep, UA, RVP, Flu, Covid-19
- Blood Cultures (peripheral culture and all lumens of central line)
- IV or IM ampicillin 75mg/kg (max dose: 2000mg) within 60 minutes**

(DO NOT WAIT FOR CXR)

↓

Admission:

-Discuss with hematology

-Consider PICU for increased WOB, fatigue, increased O_2 requirement, need for full exchange transfusion, Hgb drop

Consider diagnostic codes: sickle cell disease with acute chest syndrome

Inpatient Status

*Use a broad definition for ACS, consisting of lower respiratory symptoms including hypoxia OR a new infiltrate on CXR. Diagnosis of a clinically mild case of ACS should prompt admission for close monitoring and escalation of treatment as needed.

**If PCN allergy use levoFLOXacin 10 mg/kg/dose IV ONCE (for patients ≥ 6 months to <5 yrs, max dose: 375 mg/dose; ≥ 5 yr max dose: 750 mg/dose)

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Acute Chest Syndrome (ACS) Algorithmic Pathway (Inpatient)

ANTIBIOTICS

Ampicillin AND macrolide (*Azithromycin*) **

- 7-day total course of ampicillin ***
  5-day course of azithromycin
- Consider adding *Vancomycin* if severely ill and/or for pleural empyema or moderate or large effusion.
- (MRSA coverage)
- When treating ACS, antibiotic coverage should continue for full course even if viral source found on respiratory panel

RESPIRATORY SUPPORT

Monitoring parameters: RR, degree of air movement on auscultation, accessory muscle use, mental status, color/perfusion, O_2 sat

- O_2 as needed to maintain O_2 sat >93%
- Incentive spirometry q1 hour while awake (bubbles, pinwheel, Acapella)—***OBSERVED!***
- OOB as much as possible, PT/OT as needed
- Inhaled bronchodilators (a*lbuterol*), *scheduled*, in patients with known history of reactive airway disease/ prior ACS
  - Consider scheduled or PRN bronchodilators for non-reactive airway disease patients depending on clinical presentation
  - Short course of steroids may be needed in asthma exacerbation
- Escalating O_2 support may include: CPAP, BiPAP, intubation with mechanical ventilation or ECMO

FLUIDS

- Correct dehydration on presentation with normal saline to decrease sickling
- IVF= max ¾ x Maintenance

PAIN CONTROL

- Ketorolac (avoid concomitant oral NSAID)- see sickle cell pain guidelines for dosing
- Opioid pain meds as needed, careful to avoid sedation which may lead to hypoventilation
- Sickle Cell Disease Pain Crisis Clinical Pathway

TRANSFUSION

- ***Simple transfusion indications***
  - Improve oxygenation if O_2 sat <93%
  - Hgb drop > 1 g/dL from baseline or persistently dropping through admission and less than 10 g/dL
  - Clinical progression of disease but not impending respiratory failure

  Max hemoglobin post-transfusion 11 g/dL
- ***Exchange transfusion indications***
  - Progression of symptoms despite simple transfusion/ hgb >/= 10 g/dL
  - Severe hypoxemia
  - Multi-lobar disease
  - Previous h/o severe ACS or cardiopulmonary disease

  GOAL: decrease Hgb S to less than 30%, do not exceed Hgb of 10 g/dL

Refer to Sickle Cell Disease Blood Transfusion Pathway

Nursing Team should reference SCD Acute Chest Syndrome Nursing Standard of Care

**Use levoFLOXacin as monotherapy if allergic to penicillin or macrolides

***If discharged prior to completing 7-day antibiotic course, may send home on oral high dose Amoxicillin

Acute Chest Syndrome (ACS)

Labs/Imaging: Infections, including pneumonia, are a significant cause of ACS, so blood cultures (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 Ampicillin, Azithromycin should be administered to cover Mycoplasma (Evidence Low, consensus national panel of experts). Consider discontinuation of azithromycin if respiratory pathogen panel is negative for *Mycoplasma pneumoniae* and *Chlamydia pneumoniae.* If the patient has a penicillin allergy or a macrolide allergy, give levoFLOXacin. Consider adding vancomycin if patient severely ill and/or for pleural empyema or moderate or large effusion (see antibiotic dosing below).

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 93% (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).

Antibiotic	Dose	Comments/Considerations
Ampicillin	50 mg/kg/dose IV q6h for *7 days* (Max dose: 2000 mg/dose)	May switch to PO amoxicillin if being discharged prior to end of 7-day course
Azithromycin	10 mg/kg/dose PO once on Day 1 (max dose 500 mg/dose) *followed by* 5 mg/kg/dose PO daily on days 2 to 5 (max dose 250 mg/dose)	Consider discontinuation of azithromycin if respiratory pathogen panel is negative for *Mycoplasma pneumoniae* and *Chlamydia pneumoniae*
LevoFLOXacin	Infants ≥6 months and Children <5 years: • 10 mg/kg/dose PO/IV q12h (max 375 mg/dose) for *7 days* Children ≥5 years: • 10 mg/kg/dose PO/IV daily (max 750 mg/dose) *7 days*	Use if documented allergy to penicillin or macrolide
Vancomycin	Order via JHH-ACH Pediatric Vancomycin Focused" order set for appropriate dosing	Consider adding if patient severely ill and/or for pleural empyema or moderate or large effusion.
Amoxicillin	30 mg/kg/dose PO TID to complete *7 total days of therapy*	Use as PO option if patient is discharged prior to end of 7-day course of ampicillin

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 Clinical Pathway

SIGNS AND SYMPTOMS:

- Enlarged/ tender spleen
- Pallor
- Jaundice
- Tachycardia
- Abdominal pain/distention
- Decreased Hgb by ≥ 2 g/dL or more from baseline (+/- thrombocytopenia)

Workup and Initial Management

- Start IV, CBC, CMP, Retic, Type & Screen
- Pain management as per Sickle Cell Pain Crisis Clinical Pathway
- IVF at maintenance rate
- Consider PRBC transfusion, clarify transfusion orders with hematology (5ml/kg MAX)
- Consult Hematology re: admission
- Dx code: Hb-SS with splenic sequestration

Ongoing Management

- Continue pain management and IVF @ maintenance
- Consult surgery for possible splenectomy once no longer acutely ill. (Discuss with SCD team about timing of referral)
- Ensure patients are fully vaccinated (PPSV23 and Menveo) prior to discharge, especially in pts >/=2yrs of age if considering splenectomy
- Follow- up in clinic in 2-3 weeks for lab check and possible start of monthly PRBC transfusions

PRBC TRANSFUSION

- LOW AND SLOW
- Start with 2-5 ml/kg* over 4 hours (in general if hgb is 2 or 3 would start at 2 or 3 ml/kg respectively)
- Recheck CBC >/=2hrs post infusion and prior to ordering additional blood
- Goal hgb is >7 or hemodynamically stable
    - Risk of auto transfusion when spleen releases trapped cells, resulting in hyperviscocity

Nursing Team should reference SCD Splenic Sequestration Nursing Standard of Care

*Each 5 mL/kg PRBC transfusion should raise hemoglobin by ~1 g/dL

**PRBC are non-irradiated, sickle negative for SCD patients, can indicate sickle cell protocol in order if desired

Splenic Sequestration Algorithmic Pathway

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, thrombocytopenia or pallor. 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).

PRBC transfusion for splenic sequestration: Start with 2-5 ml/kg over 4 hours (in general if hgb is 2 or 3 would start at 2 or 3 ml/kg respectively). Recheck CBC ≥2hrs post infusion and prior to ordering additional blood. Goal hgb is >7 g/dL or hemodynamically stable. You do not need to transfuse patient back to their baseline hemoglobin. Risk of auto transfusion when spleen releases trapped cells, resulting in hyperviscocity and increased risk of stroke.

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Sickle Cell Disease Blood Transfusion Clinical Pathway

SIMPLE TRANSFUSION INDICATIONS

- Improve oxygenation if O_2 sat <93%
- Hgb drop > 1 g/dL from baseline or persistently dropping through admission and less than 10 g/dL
- Clinical progression of disease but not impending respiratory failure

Max hemoglobin post-transfusion 11 g/dL

EXCHANGE TRANSFUSION INDICATIONS

- Progression of symptoms despite simple transfusion/ hgb ≥ 10 g/dL
- Severe hypoxemia
- Multi-lobar disease
- Previous h/o severe ACS or cardiopulmonary disease

GOAL: decrease Hgb S to less than 30%, do not exceed Hgb of 10 g/dL

Consult hematology prior to ordering blood
M-F 8am-5pm: Notify Sickle Cell Team also

Able to obtain appropriate access (Two 18 g PIV or central lines) and ICU/ One Blood availability?

YES

One Blood Orders for Depletion Exchange

- Call One Blood to request inpatient depletion exchange (727-568-1111)
- **One Blood RBC Pheresis- ACH Therapy Plan** in Epic
- Minimum HCT 21, Goal HCT 30, Fraction of Cells Remaining (FCR) 25 **
- One Blood will give the number of units needed for goal
- Post hemoglobin electrophoresis collected after exchange transfusion. Goal Post Hgb S < 20%

NO

Partial Exchange (Therapeutic Phlebotomy followed by Simple Transfusion)

- **Epic order set: JHH pediatric partial exchange transfusion**
- If patient is too small for depletion exchange **and Hgb > 7**, partial exchange is preferred to simple transfusion. **Max Goal Hgb 10 g/dL**. *
- Amount of PRBC to order/ phlebotomize:
  - Hgb 7-8 g/dl, Phleb 5 ml/kg prior to transfusing 15 ml/kg.
  - Hgb 8-9 g/dL, Phleb 10 ml/kg prior to transfusing 15 ml/kg.
  - Hgb > 9, Phleb 15 ml/kg prior to transfusing 15 ml/kg (do not transfuse more than phlebotomy).

ONLY to be used when meets criteria for exchange transfusion and unable to do full depletion (due to patient being too small/ unable to get appropriate access/ ICU)

*Each 5 mL/kg PRBC transfusion should raise hemoglobin by ~1 g/dL

**PRBC are non-irradiated, sickle negative for SCD patients, can indicate sickle cell protocol in order if desired

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Sickle Cell Disease Stroke Algorithmic Pathway

Concern for Stroke:

Abnormal neurologic exam, new onset seizure activity, aphasia, altered mental status, etc.

Refer to JHACH Childhood Arterial Ischemic Stroke Clinical Pathway

- STAT Brain MRI w/wo contrast if available, if not STAT Head CT w/o contrast
- Check labs: CBC, retic, CMP, type and screen, hemoglobin electrophoresis STAT
- Discuss with Neurology re: assessment
- Discuss with Hematology re: simple, partial or depletion exchange transfusion
- Admit to PICU under inpatient setting
- Call One Blood for emergent inpatient depletion exchange (727-568-1111)

↓

Exchange Transfusion

- Hemoglobin electrophoresis collected before and after exchange transfusion. Goal Post Hgb S < 20%
- Brain Angio MRA w/ MRV to be done after exchange

↓

Ongoing Care

- Surgery consult for **vortex port** placement for ongoing monthly depletion exchanges
- Set up with Rehab therapy, inpatient vs outpatient
- Neurology follow-up
- Neuro-psych evaluation

*Each 5 mL/kg PRBC transfusion should raise hemoglobin by ~1 g/dL

**PRBC are non-irradiated, sickle cell negative for SCD patients, can indicate sickle cell protocol in order if desired

Sickle Cell Disease Stroke Algorithmic Pathway

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.

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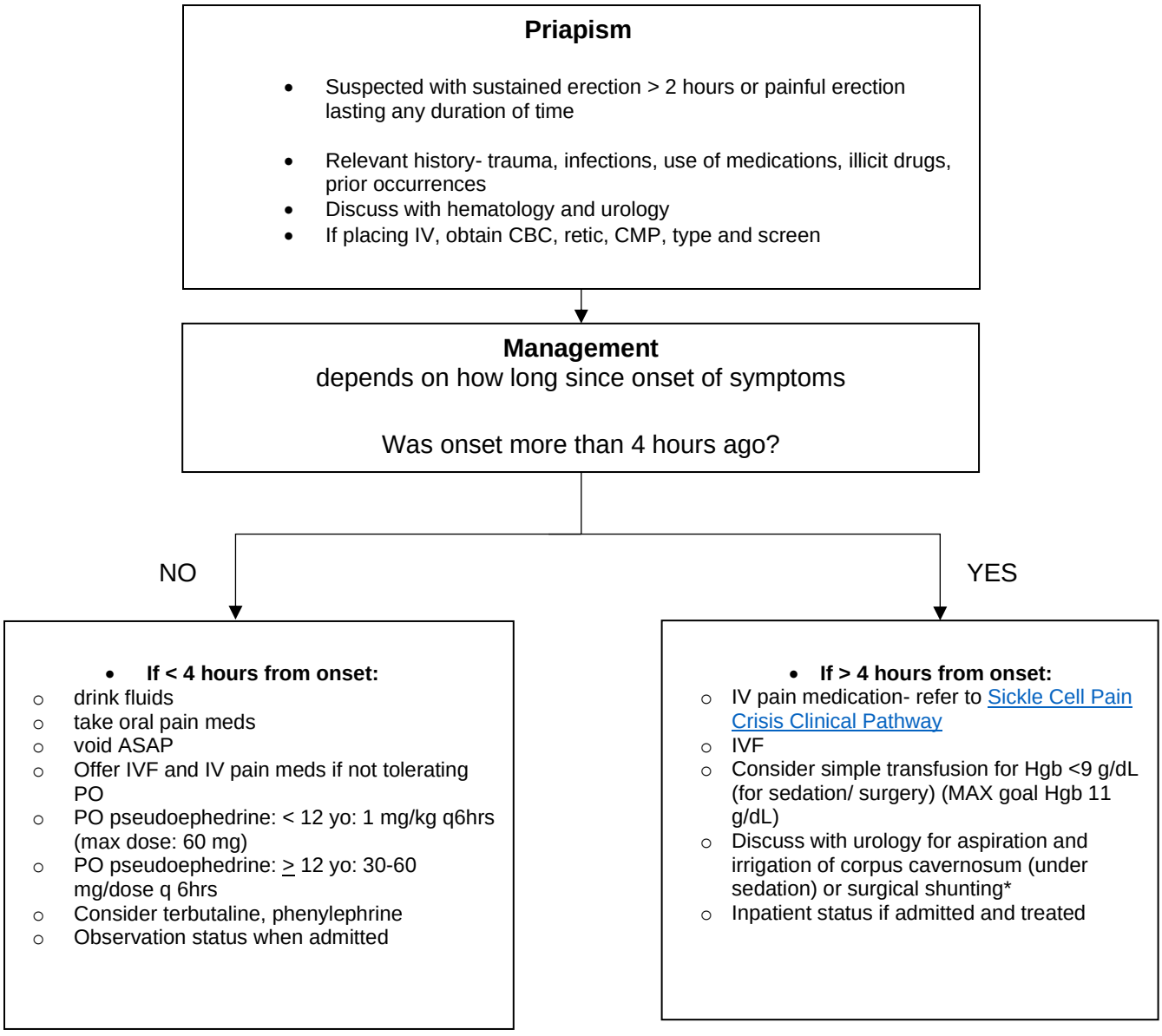
Sickle Cell Disease Priapism Clinical Pathway



* If sedation or surgical shunting is needed, see Sickle Cell Disease Surgery/ Anesthesia Recommendations

Sickle Cell Disease Priapism Algorithmic Pathway

Labs/Imaging: CBC, CMP, Reticulocyte count, type and screen 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 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 consult (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

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Sickle Cell Disease Cholelithiasis/Cholecystitis Clinical Pathway

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 mL/kg NS) and start MIVF
- IV pain medication- reference Sickle Cell Pain Crisis Clinical Pathway
- Give IV ondansetron as needed for N/V
- RUQ ultrasound
- If febrile, obtain blood cultures peripherally and all lumens of central line (if present), start IV cefTRIAXone 50 mg/kg (max dose: 2000 mg) ASAP; Consider piperacillin/tazobactam 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 opioid 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.

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Sickle Cell Disease Aplastic Crisis Clinical Pathway

Signs and Symptoms:

- Jaundice
- Tachycardia
- Decreased Hgb by ≥2g/dL or more from baseline (+/- thrombocytopenia/leukopenia) without appropriate retic response

Workup and Management

- Start IV, CBC, CMP, Retic, Type & Screen
- Send viral panel including Parvo B19 and EBV
- Pain management per Sickle Cell Pain Crisis Clinical Pathway
- IVF@ maintenance rate
- Consider PRBC transfusion
- Dx code: Aplastic crisis

Blood Transfusion

- Hgb decreased by ≥ 2 gm/dl **AND** inappropriate retic response
- Transfuse to goal of 8-9 g/dL*
- Discharge home when clinically stable with close follow up - discuss timeframe with outpatient sickle cell team

Nursing Team should reference SCD Aplastic Crisis Nursing Standard of Care

*Each 5 mL/kg PRBC transfusion should raise hemoglobin by ~1 g/dL

**PRBC are non-irradiated for SCD patients, can indicate sickle cell protocol in order if desired

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Sickle Cell Disease Surgery/ Anesthesia Recommendations

Transfusion and Pre-op/ Sedation Guidelines

-Check CBC 1-3 days prior (approx.) to surgery
-PRBC transfusion with goal hemoglobin 9-10 g/dL (MAX 11 g/dL) *
-Admit to hematology service day prior to surgery for PRBC transfusion**
-After transfusion complete, start IVF at maintenance rate overnight until surgery

Surveillance studies (transcranial doppler ultrasound, routine labs) should be up to date to provide surgical clearance

Intra-op and Post-op Guidelines

-O_2 to keep saturations >93% during surgery with wean while in PACU if possible
-Continue MIVF through surgery until adequate PO post-op
-Pain control post-op to avoid splinting
-Incentive spirometry (bubbles, pinwheel, Acapella) q1hr (or more frequent) while awake
-OOB and resume ADLs as soon as possible

-Possible **discharge home** on day of surgery for dental procedure, radiology procedure, T&A
-**Admit** at least overnight for abdominal or complicated surgery AND/OR history of acute chest syndrome/O_2 requirement, moderate/severe pulmonary disease, or previous surgery complication

Dental Procedures

In-office
-Do not use Nitrous Oxide
-Lidocaine local injection may be used

Operating Room with general anesthesia
-Admit for IVF hydration +/- PRBC transfusion
-Discharge on day of surgery if pain well controlled and taking adequate PO

Antibiotic prophylaxis on case-by-case basis (ex: Amoxicillin)

Sedated Radiology Scans

-Check CBC 1-3 days prior (approx.)
-No transfusion PRBC if Hgb >8.5 g/dL
-IVF hydration at maintenance while admitted prior to sedation
-Careful consideration for use of contrast depending on renal function
-Discharge home after anesthesia recovery

*Each 5 mL/kg PRBC transfusion should raise hemoglobin by ~1 g/dL
**PRBC are non-irradiated, sickle negative for SCD patients, can indicate sickle cell protocol in order if desired

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

1) Kavanaugh PL, Sprinz PG, Wolfgang Tl, et al. Improving the management of vaso-occlusive episodes in the pediatric emergency department. *Pediatrics*. 2015 Oct;136(4):e1016-25. Epub 2015 Sep 21 (Evidence low)
2) Treadwell MJ, Bell M, Leibovich Sa, et al. A quality improvement initiative to improve emergency department care for pediatric patients with sickle cell disease. *J Clin Outcomes Manag*. 2014 Feb;21(2):62-70 (Evidence low)
3) Lin SM, Strousse JJ, Whiteman LN et al. Improving Quality of Care for Sickle Cell Patients in the Pediatric Emergency Department. *Ped Emer Care*. 2016 Jan;32(1):14-16 (Evidence low)
4) Mathias MD, McCavitt. Timing of opioid administration as a quality indicator for pain crises in sickle cell disease. *Pediatrics*. 2015 Mar;123(3):475-82 (Evidence low)
5) Morrissey LK, Shea JO, Kalish LA, et al. Clinical practice guideline improves the treatment of sickle cell disease vasoocclusive pain. *Pediatr Blood Cancer*. 2009;52(3):369 (Evidence low)
6) Jayaran A, Nagel RW, Jasty R. Impact of a clinical pathway on quality of care in sickle cell patients. J *Pediatr Hematol Oncol*. 2010;32(7):537 (Evidence low)
7) Ender KL, Krajewski JA, Babineau J, et al. Use of a clinical pathway to improve the acute management of vaso-occlusive crisis pain in pediatric sickle cell disease. *Pediatr Blood Cancer*. 2014 Apr;61(4):693-6 (Evidence low)
8) Krishnamurti L, Smith-Packard B, Gupta A, et al. Impact of individualized pain plan on the emergency management of children with sickle cell disease. *Pediatr Blood Cancer*. 2014 Oct;61(10):1747-53 (Evidence low)
9) Frei-Jones MJ, Baxter AL, Rogers ZR, Buchanan GR. Vaso-occlusive episodes in older children with sickle cell disease: emergency department management and pain assessment. *J Pediatr*. 2008 Feb;152(2):281-5 (Evidence low)
10) Morris C, Vichinsky E, Styles L. Clinician Assessment for Acute Chest Syndrome in Febrile Patients with Sickle Cell Disease: Is It Accurate Enough? *Annals of Emergency Medicine.* 1999 July; 34 (1): 64-9
11) National Guideline Clearinghouse (NGC). Guideline summary: Sickle cell acute painful episode: management of an acute painful sickle cell episode in hospital. In: National Guideline Clearinghouse (NGC) [Web site]. Rockville (MD): Agency for Healthcare Research and Quality (AHRQ); 2012 Jun 01. [cited 2017 Jan 13]. Available: https://www.guideline.gov (Evidence low)
12) National Guideline Clearinghouse (NGC). Guideline summary: Managing acute complications of sickle cell disease. In: Evidence-based management of sickle cell disease. In: National Guideline Clearinghouse (NGC) [Web site]. Rockville (MD): Agency for Healthcare Research and Quality (AHRQ); 2014 Jan 01. [cited 2017 Jan 13]. Available: https://www.guideline.gov (Evidence low)
13) Debaun M, Vichinsky E, Mahoney D, Schrier S, Tirnauer J. Vasoocclusive pain management in sickle

cell disease. *UpToDate*. Aug 23, 2016 (Evidence low)
14) Beiter JL, Simon HK, Chambliss CR, et al. Intravenous ketorolac in emergency department management of sickle cell pain and predictors of its effectiveness. *Arch Pediatr Adolesc.* Med 2001; 155(4):496 (Evidence low)
15) Murphy A, O'sullivan R, Wakai, et al. Intranasal fentanyl for the management of the acute pain in children. *Cochrane Database Syst Rev*. 2014 (Evidence low)
16) Rogers Z, Mahoney D, Schrier S, Tirnauer J. Management of fever in sickle cell disease. *UpToDate*. Dec 16, 2016 (Evidence low)
17) Baskin MN, Goh XL, Heeney MM, et al. Bacteremia risk and outpatient management of febrile patients with sickle cell disease. *Pediatrics*. 2013; 131:1035 (Evidence low)
18) Bansil NH, Kim TY, Tieu L, Barcega B. Incidence of serious bacterial infections in febrile children with sickle cell disease. *Clin Pediatr*. 2013 Jul;52(7):661-6 (Evidence low)
19) Heeney M, Mahoney D, Mallory G, Schrier S, Tirnauer J. The acute chest syndrome in children and adolescents with sickle cell disease. *UpToDate*. Dec 9, 2015 (Evidence low)
20) U.S. Department of Health and Human Services, National Institutes of Health, National Heart Lung and Blood Institute. (2014). Evidence based management of sickle cell disease, expert panel report 2014.
21) Smith-Whitley K, Zhao H, Hodinka RL, et al. Epidemiology of human parvovirus B19 in children with sickle cell disease. *Blood* 2004; 103;422.
22) Schultz CL, Tchume-Johnson T, Schapira MM, Bellamy S, Smith-Whitley K, Ellison A. Adherence to prompt fever evaluation in children with sickle cell disease and the health belief model. *Pediatric Blood cancer.* 2015 Nov; 62(11):1968-73. Epub 2015 July 14.
23) Narang S, Fernandez ID, Chin N, Lerner N, Weinberg GA. Bacteremia in children with sickle hemoglobinopathies. *J Pediatr Hematol Oncol*. 2012;34(1):13.
24) National Institutes for Health and Clinical Excellence. (2012) Sickle cell acute painful episode: management of an acute sickle cell episode in hospital. http://www.nice.org/uk/nicemedia/live/13772/59765/59765.pdf
25) Wang, J.C.; Kavanagh, P.L.; Little, A.A.; Holliman, J.B.; & Sprinz, P.G. (2011) Quality-of-care indicators for children with sickle cell disease. *Journal of the American Academy of Pediatrics*, 128 (484).

Clinical Pathway Team

Sickle Cell Disease Clinical Pathway

Johns Hopkins All Children's Hospital

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Disclaimer

Clinical Pathways are intended to assist physicians, physician assistants, nurse practitioners and other health care providers in clinical decision-making by describing a range of generally acceptable approaches for the diagnosis, management, or prevention of specific diseases or conditions. The ultimate judgment regarding care of a particular patient must be made by the physician in light of the individual circumstances presented by the patient.

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APPENDIX A:
SICKLE CELL ACUTE CHEST SYNDROME (ACS) NURSING STANDARD OF CARE

Nursing Standard of Care

The patient will experience appropriate oxygenation and minimal side effects from a lower respiratory tract infection.

Patient Outcomes

The patient will:

- Be medically stable throughout periods of ACS
- The patient's onset of ACS will be identified as soon as the presence of defining signs and symptoms appear.
- The patient will maintain adequate oxygenation throughout period of ACS.
- The patient will receive prescribed antimicrobials in a timely manner to limit course of ACS.
- Perform self-care activities to prevent complications from acute chest syndrome.
- The patient/family will demonstrate/verbalize an understanding of activities, daily care and measures to prevent worsening of infection.
- The patient/family will verbalize signs and symptoms of infection.
- The patient will perform incentive spirometry 10 breaths every 1 hour while awake.
- The patient should ambulate at least 4 times daily and stay out of bed for at least 2 hours each time to encourage lung expansion.

Standard of Practice

Interventions:

The nurse will:

- Be knowledgeable regarding definition of acute chest syndrome (ACS): an acute illness characterized by a new pulmonary infiltrate on chest x-ray and one or more of the following symptoms:
    - Lower respiratory symptoms (dyspnea, respiratory distress, cough)
    - Fever
    - Chest pain
    - Hypoxemia.
- Additional symptoms of ACS may be abdominal pain (especially in small children) and back pain. Monitor all sickle cell patients for signs and symptoms of ACS understanding that the first sign may be decreased breath sounds and a cough.
- Assess patient's breath sounds and work of breathing every 4 hours understanding shortness of breath, tachypnea, retractions and wheezing are signs and symptoms associated with ACS. Notify the MD/NP/PA of any changes in patient status.
- Monitor the patient's oxygen saturation level by continuous pulse oximetry.
- Administer oxygen as ordered by nasal cannula or face mask if O2 saturation by pulse oximeter is below 93% or if there is a 3 % decrease in the patient's baseline oxygen level. The MD/NP/PA will communicate the parameter for notification if differing from 93%.
- Anticipate that a finding of hypertension in the setting of ACS is a risk factor for PRES syndrome (Posterior reversible encephalopathy syndrome).
- Encourage patient to perform Incentive Spirometer or age-appropriate pulmonary exercise 10x every hour; Q1 hour while awake and document.
- Make sure patient is out of bed for 1-2 hours with a goal of 4 times a day, for a minimum of 6 hours a day.
- Encourage patient to ambulate at least 4 times a day.
- Anticipate IV antibiotic order and to administer within 1 hour of arrival/admission-
- *Contact MD/NP/PA STAT if sepsis is suspected
- Anticipate IV fluid administration at ¾ maintenance rate.
- Monitor CBC for acute anemia and administer blood products as ordered. Patient may have increased oxygen demand or decrease in hemoglobin concentration.
- Make sure Oxygen saturations are documented every 4 hours along with routine vital signs.
- Ensure patient's pain level is controlled following the Sickle Cell SOC for pain assessment and medication administration.

APPENDIX B:
SICKLE CELL APLASTIC CRISIS NURSING STANDARD OF CARE

Nursing Standard of Care

The patient will experience minimal side effects from aplastic crisis.

Patient Outcomes

The patient will be medically stable throughout periods of aplastic crisis.

Standard of Practice

Interventions:
The nurse will:

1. Be knowledgeable regarding the definition of aplastic crisis: a transient failure of RBC production primarily caused by parvovirus B19 infection.

2. Monitor all sickle cell patients for signs and symptoms of aplastic crisis including: lethargy, tachypnea, tachycardia, fever, low hemoglobin and reticulocyte count (usually < 1%); with the understanding that the first sign may be the presence of a nonspecific viral illness.

3. Monitor lab work and notify NP/MD/PA of abnormal lab values, especially a drop in hemoglobin concentration or a hemoglobin level of 2 g/dL or more below the patient's baseline level or a drop in the reticulocyte count.

4. Recognize the significance of aplastic crisis in sickle cell patients because the lifespan of their red blood cells is significantly shorter than the healthy red blood cells (20-40 days v. 120 days). Administer PRBCs as ordered. If more than 15 ml/kg is ordered, please clarify with the ordering provider.

5. Contact MD/NP/PA for isolation order if parvovirus is suspected, educating patient/family about isolation guidelines. *Provide proper PPE instruction

6. Expect the duration of the crisis to last approximately 7-10 days.

APPENDIX C:
SICKLE CELL FEVER NURSING STANDARD OF CARE

Nursing Standard of Care

The patient will become afebrile.

Patient Outcomes

The patient will be medically stable throughout periods of fever.

Standard of Practice

Interventions:
The nurse will:

- Recognize fever as a medical emergency.
- Be knowledgeable regarding the criteria for a fever in a patient with sickle cell is >/= 38.3 C (101 F)
- Monitor all sickle cell patients for signs and symptoms of complications from fever including: acute chest syndrome, aplastic crisis, pain crisis, and splenic sequestration.
- Be knowledgeable about administration of antipyretics such as acetaminophen and ibuprofen. No antipyretic should be given before patient is seen and fever is documented.
- Clarify PRN orders for Ibuprofen and Tylenol if both are ordered.
- Contact MD/NP/PA for isolation order if parvovirus is suspected, educating patient/family about isolation guidelines.
- Collect blood and urine cultures, CBC with reticulocyte count, chemistries, and UA as ordered. Anticipate chest x-ray.
- Anticipate IV antibiotic order (**administer within 1 hour of arrival/admission)** and IVF (anticipate ¾ -1 times maintenance rate- depending on other symptoms: see acute chest and pain standards of care) as ordered, recognizing that prophylactic penicillin may be held while patient is receiving other antibiotics, but should be restarted at discharge or after completion of other antibiotics.
- Notify provider if IV access is unable to be obtained within 30 minutes of arrival so IM antibiotic can be considered.
- Encourage patient to perform Incentive Spirometer or age-appropriate pulmonary exercise 10x every hour; Q1 hour while awake and document.
- Make sure patient is out of bed for 1-2 hours with a goal of 4 times a day, for a minimum of 6 hours a day.
- Encourage patient to ambulate at least 4 times a day.

APPENDIX D:
SICKLE CELL PAIN NURSING STANDARD OF CARE

Nursing Standard of Care

The patient's experience of pain will be minimal and limited to a level acceptable to the patient.

Patient Outcomes

The patient will experience an optimal level of pain relief and can expect his/her report of pain to be accepted, assessed and intervention taken.

Standard of Practice

Interventions:
The nurse will:

- Monitor all sickle cell patients for signs and symptoms of pain and document every 4 hours or PRN using the proper pain scale.
- Recognize the specific differences in causation, experience and treatment of pain in the sickle cell patient population.
- Make appropriate referrals, including: Child Life, Rehab Services, Social Work, etc.
- Upon initial presentation with an acute pain episode reassess for pain 30 minutes after administration of the first pain medication dose.
- Notify MD/NP/PA after 30-minutes after assessment and anticipate the order for additional pain medications.
- Administer pain medications as ordered; anticipating order for PCA.
- Administer appropriate non-pharmacologic pain interventions such as heat packs, warm baths, and warm blankets as needed.
- Administer IVF (anticipate 1-1 ½ times maintenance) as ordered; recognizing that often pain in patients with sickle cell can be caused by dehydration.
- Be knowledgeable about specific pain medications for patients with sickle cell, including using ketorolac for a maximum of 5/30 days a month, verify with pharmacy.
- Anticipate gastric-protective agent, order while on ketorolac.
- Be knowledgeable about appropriate supportive care for patients receiving opioids (i.e., discuss with MD/NP/PA need for stool softeners, laxatives, antipruritic, anti-nausea medications).
- Educate patient and family about the use of pain medications for treatment of pain.
- Encourage patient to perform Incentive Spirometer or age-appropriate pulmonary exercise 10x every hour; Q1 hour while awake and document.
- Make sure patient is out of bed for 1-2 hours with a goal of 4 times a day, for a minimum of 6 hours a day.
- Encourage patient to ambulate at least 4 times a day.
- Make sure Oxygen saturations are documented every 4 hours along with routine vital signs **and** **monitor continuous O2 saturations while on a PCA basal rate.**
- **Notify MD/NP/PA if patient O2 sat is <93% or 3% less than baseline.**

APPENDIX E:
SICKLE CELL SPLENIC SEQUESTRATION NURSING STANDARD OF CARE

Nursing Standard of Care

The patient will experience appropriate hemodynamic stability and minimal side effects from splenic sequestration.

Patient Outcomes

The patient will be medically stable throughout periods of splenic sequestration.

a. The patient's onset of splenic sequestration will be identified as soon as the presence of defining signs and symptoms appear.
b. The patient will maintain adequate blood pressure, oxygenation and level of consciousness during time of splenic sequestration.

Standard of Practice

Interventions:
The nurse will:

- Recognize splenic sequestration as a medical emergency.
- Anticipate continuous cardiac and pulse oximetry monitoring.
- Be knowledgeable regarding the definition of splenic sequestration including: a sudden enlargement of the spleen greater than 2 cm above baseline caused by intrasplenic trapping of blood which results in a precipitous decrease in the hemoglobin level and platelet count and increased reticulocyte count.
- Monitor all sickle cell patients, especially those with recent bacterial or viral infections, for signs and symptoms of splenic sequestration including: pallor, lethargy, lack of energy, loss of appetite, tachycardia, tachypnea, abdominal fullness and, in toddlers, often fussiness and irritability.
- Understand splenic sequestration may occur as early as one month of age but it is more common in children between the ages of 1 and 4 years of age but can happen at any age.
- Monitor the patient's hemoglobin level; recognizing that a decrease 2g/dL from normal hemoglobin concentration can be an indication of splenic sequestration.
- Confirm with blood bank the patient has a current type and screen.
- Understand if a patient has had one splenic sequestration, they are at an increased risk of having another.
- Assess the patient for an enlarged spleen in the left upper quadrant, using caution with palpation.
- The MD/NP/PA using a marker will document the spleen border on the abdomen to determine an increase or decrease in spleen size.
- Understand that splenic sequestration can often be related to an episode of acute chest syndrome and is a major cause of acute anemia. Administer oxygen as ordered if the O2 saturation is below 93% or if there is a 3 % decrease in the patient's baseline oxygen level. The MD/NP/PA will communicate the parameter for notification if differing from 93%.
- Monitor the patient's blood pressure understanding that patients with splenic sequestration are at risk for hypovolemic shock.
- Anticipate orders for IV fluids and PRBCs noting the transfusion is aimed at a **partial correction **** Excessive transfusion should be avoided as the patient may experience a release of PRBCs from the spleen causing an unexpected rise in hemoglobin.
- Call MD if order for PRBCs volume is greater than 5 ml/kg.
- Ensure the patient's pain level is controlled adequately. Refer to Sickle Cell Pain SOC.
- Recognize parent teaching by the team includes proper palpation of the spleen and determine baseline spleen for their child.
- Recognize parent teaching should include signs and symptoms of acute anemia including: pallor, lethargy, lack of energy, loss of appetite, tachycardia, tachypnea, and in toddlers, often fussiness and irritability.