**CLINICAL PATHWAY: Fever in a Patient with Sickle Cell Disease**

**Inclusion Criteria:**
- ≥ 2 months of age with sickle cell disease (HgbS, HgbSC, HgbS beta thal) and temp ≥101°F (38.5°C)

**Exclusion Criteria:**
- ≤ 2 months old, sickle cell trait, signs of sepsis (see Septic Shock Pathway), clinical suspicion for Multi-System Inflammatory Syndrome in Children (see MIS-C Clinical Pathway)

### If presents to ED: Triage Level 2

#### RN Evaluation:
- Vital signs, continuous pulse ox
- Blood culture (from all lumens of CVLs)
- If no CVL, obtain peripheral culture
- CBC & reticulocyte count & STAT procalcitonin
- Give acetaminophen 15 mg/kg/dose q6hr (max 1000 mg/dose; max 75 mg/kg/day, not to exceed 4000 mg/day) if not received in past 4 hours and/or
  - Ibuprofen 10 mg/kg/dose q6hr (max 800 mg/dose).
  - Toradol IV 0.5 mg/kg/dose (max 30 mg/dose) q6hr, if not received in past 6 hours
- STAT:
  - If patient with persistent fever
    - CBC & reticulocyte count & STAT procalcitonin q48hr (or sooner, if clinically indicated)
  - If ill appearing:
    - Ceftriaxone IV
- Note: Patients with severe sickle cell disease completed.
  - Penicillin prophylaxis. If patient is on prophylaxis, can pause prophylaxis while on antibiotics above. Resume prophylaxis once antibiotic therapy is completed.
- If cephalosporin allergy:
  - Add vancomycin IV
  - Discharge after antibiotics administered:
    - If ceftriaxone given prior discharge: no additional antibiotics needed
    - If received levofloxacin ≤ 1 dose prior to discharge: give prescription for 2nd dose 12 hours later (see above for dosing – IV and PO dosing are equal)
    - Continue penicillin prophylaxis (if taking)
    - Outpatient follow up plan discussed with on-call Heme/Onc attending

### Admission Criteria:
- ≤ 12 months old
- Hx of encapsulated bacteria/sepsis
- WBC < 5,000 or > 30,000
- Platelet < 100,000
- Ill appearing
- Oxygen requirement
- Hgb < 6 g/dL or 2 g/dL below baseline
- Hypotension
- Poor perfusion
- New infiltrate on CXR
- Dehydration
- Concern for caregiver ability to care for patient

### Provider Evaluation:
- Consider further diagnostic work-up based upon history and physical exam
- If concern for acute chest syndrome:
  - Call Heme/Onc to discuss all patients
  - Pneumococcal vaccine not routinely indicated

### Discharge criteria:
- Well-appearing and tolerating PO; negative blood cultures; outpatient follow up in place

### Consults:
- Call Heme/Onc to discuss all patients

### Inpatient Care
- CBC & reticulocyte count & STAT procalcitonin q48hr (or sooner, if clinically indicated)
- If patient with persistent fever: blood cultures from all CVL lumens or peripheral blood culture q24hr

### PMA (Post-Menstrual Age) = gestational age + postnatal age

### Antibiotics:
- Ceftriaxone 75 mg/kg IV (max 2 g/dose)
- If cephalosporin allergy: levofloxacin IV
  - ≤ 5 years old: 10 mg/kg/dose q12hr
  - ≥ 5 years old: 10 mg/kg/dose daily (max 750 mg/day)
- If ill appearing:
  - Vancomycin IV: ≤ 3 months old: 10 mg/kg/dose q12hr or as determined by pharmacy based on estimated AUC; 6-11 months old: 70 mg/kg/day div q12hr; ≥ 12 years old: 60 mg/kg/day div q12hr
  - If concern for acute chest syndrome: add azithromycin 10 mg/kg on day 1 (max 500 mg/dose), then 5 mg/kg once daily on day 2-5 (max 250 mg/dose). If respiratory BIOFIRE was sent and negative for atypical organisms, discontinue azithromycin.
- If presents to ED:
  - Hold purple top for Type & Screen, green top for BMP or LFT
  - CRP, chemistry, LFTs, Type & Screen, urinalysis, CXR (if concern for acute chest syndrome); respiratory BIOFIRE not routinely indicated
  - If no CVL, obtain peripheral culture
  - If source of infection identified, treat appropriately. Otherwise, continue antibiotics below.

### Discharge after antibiotics administered:
- If ceftriaxone given prior discharge: no additional antibiotics needed
- If received levofloxacin ≤ 1 dose prior to discharge: give prescription for 2nd dose 12 hours later (see above for dosing – IV and PO dosing are equal)
- Continue penicillin prophylaxis (if taking)
- Outpatient follow up plan discussed with on-call Heme/Onc attending

### Meets admission criteria?
- YES
- NO

### JUDGMENT.

**THIS PATHWAY SERVES AS A GUIDE AND DOES NOT REPLACE CLINICAL JUDGMENT.**

©2019 Connecticut Children’s Medical Center. All rights reserved.