CLINICAL PATHWAY:

Fever in a Patient with Sickle Cell Disease

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

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**)

Outpatient Clinic and Emergency Department Care

If presents to ED: Triage Level 2

RN Evaluation:

- Vitals, continuous pulse ox
- Blood culture (from all lumens of CVLs)
  - If no CVL, obtain peripheral culture
- CBC & Reticulocyte count & STAT procalcitonin
  - Hold purple top for Type & Screen, green top for BMP or LFT's
- 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), or **Toradol IV** 0.5 mg/kg/dose (max 30 mg/dose) q6hr, if not received in past 6 hours

Provider Evaluation:

- **STAT:** order antibiotics (see dosing below)
- Consider further diagnostic work-up based upon history and physical exam
  - CRP, chemistry, LFTs, Type & Screen, urinalysis, CXR (if concern for Acute Chest Syndrome); respiratory BIOFIRE not routinely indicated

Is patient septic AND/OR suspicion for acute chest syndrome?

YES → Proceed off-pathway. Proceed to **Septic Shock Pathway,** or manage acute chest and notify attending/fellow immediately

NO

Antibiotics:

Antibiotics should be given *within 1 hour* of presentation

If source of infection identified, treat appropriately AND give antibiotics below.

- **Ceftriaxone** 75 mg/kg IV (max 2 g/dose)
- *If Cephalosporin allergy:* **Levofloxacin IV:** 6 mo-<5 years old: 10 mg/kg/dose BID; ≥5 years old: 10 mg/kg/dose daily (max 750 mg/day)
- *If ill appearing:* add **Vancomycin IV**: <52 weeks PMA‡/about <3 mo old: 15 mg/kg q8hr or as determined by pharmacy based on estimated AUC; ≥52 weeks PMA‡/about ≥3 months old – 11 years old: 70 mg/kg/day div q6hr; ≥12 yrs old: 60 mg/kg/day div q8hr
- *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.

Consults:

- Call Heme/Onc to discuss all patients

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

[1]Admission Criteria:

- <12 months old
- Hx of encapsulated bacteremia/ 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

Meets admission criteria[1]?

NO → **Discharge after antibiotics administered**

- *If ceftriaxone given prior discharge*: no additional antibiotics needed
- *If received **Levofloxacin** x1 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

YES

Inpatient Care

Admit to Hematology/Oncology Service

If source of infection identified, treat appropriately. Otherwise, continue antibiotics below.

Antibiotics:

- **Ceftriaxone IV** 75 mg/kg/day divided q12hr (max 2 g/dose)
- *If Cephalosporin allergy:*
  - **Levofloxacin IV:** 6 mo-<5 years old: 10 mg/kg/dose BID; ≥5 years old: 10 mg/kg/dose daily (max 750 mg/day)
- *If Ill appearing:*
  - Add **Vancomycin IV**: <52 weeks PMA‡/about <3 mo old: 15 mg/kg q8hr or as determined by pharmacy based on estimated AUC; ≥52 weeks PMA‡/about ≥3 months old – 11 years old: 70 mg/kg/day div q6hr; ≥12 yrs old: 60 mg/kg/day div q8hr
  - Can discontinue if blood cultures negative x48 hours (even if still febrile)
- Note: Patients with severe sickle cell disease ≤5 yrs old (and those >5 yrs old with hx of splenectomy or invasive pneumococcal disease) should be on penicillin prophylaxis. If patient is on prophylaxis, can pause prophylaxis while on antibiotics above. Resume prophylaxis once antibiotic therapy is completed.

Lab work:

- 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

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

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