Management of fever in children with sickle cell disease

Introduction: Fever is a common presenting symptom for many manifestations of sickle cell disease (SCD). In particular, fever can frequently be the first indication of serious and life-threatening bacterial infections. As a result, patients with SCD and fever should be evaluated and treated promptly to avoid significant morbidity and mortality.

If a patient with a diagnosis of sickle cell disease comes in with fever - **triage should be expedited** and a **full set of vital signs including pulse oximetry** should be obtained.

All well-appearing* febrile sickle cell patients will follow this pathway for evaluation and management

Fever is defined as :
Temperature of ≥ 101°F (38.3°C) PO/rectal if above 6 months of age and
Temperature of > 100.4°F (38°C) rectal < 6 months of age

***If sick appearing/septic/hemodynamic instability: follow sepsis pathway**

Evaluation:

History:
In addition to routine H/P, history should also focus on
-History of Bacteremia/Sepsis particularly pneumococcus/meningococcus
-History of splenectomy
-Missing, delayed age appropriate immunizations (both Prevnar, Pneumovax, Meningococcal B)
-Non-adherence with penicillin prophylaxis
-Assess for possibility of non-adherence with outpatient follow-up

Labs:
- CBC with differential
- Reticulocyte count
- Electrolytes, renal and liver function tests (for hydration status, signs of hemolysis, organ injury)
- Blood culture
- Urine culture (if appropriate - UTI signs/symptoms, male <6 months or females <24 months).

(Consider drawing type and screen to hold/send if patient is sick appearing, pale or concerns for acute chest syndrome)

*** If patient has a port – port to be accessed and labs and blood culture to be drawn from port.**
**** Place PIV while drawing labs**

Imaging:
-CXR (low threshold for obtaining CXR as patients with sickle cell disease are at a higher risk of having an occult pneumonia)
Recommend CXR for all patients with cough, chest pain, tachypnea, hypoxia, or physical findings suggestive of pneumonia, when breath sounds cannot be auscultated throughout lungs, prior history of acute chest syndrome or less than 3 years of age

Management:

-Rapid administration of antibiotics. All effort to complete within 60 minutes of triage.
Following lab studies, patients will receive one dose of Ceftriaxone (75mg/kg) IV x1 (max. dose 2 grams).
If patient is allergic to Ceftriaxone they should receive Clindamycin 10 mg /kg IV (max. dose 900 mg) q8h and patient will be admitted.*
*Add vancomycin (15mg/kg IV q 6 hr) for suspected meningitis, hypotensive shock, suspected sepsis (follow sepsis pathway)
** Add azithromycin for acute chest syndrome (follow management guidelines for acute chest syndrome)

Supportive care
- Fluids can be started at 1 X Maintenance unless needs more for dehydration.
No IVF bolus should be given unless signs of dehydration.
-Supportive medications may be given as needed for pain control, nausea etc

Criteria for discharge after one dose of antibiotics with phone follow-up next day:

A. Clinical →

1. > 12 months of age
2. Temperature < 40°C
3. Well-appearing and tolerating PO
4. No concerns for splenic sequestration, acute chest syndrome, neurological impairment or vaso-occlusive episode requiring admission
5. No focal bone pain concerning for osteomyelitis
6. Stable on room air

B. Labs/X-Ray →

1. Hemoglobin > 5 g/dL and not less than 2g/dL below baseline. Platelets > 100K
2. WBC < 30 K/uL and > 5 K/uL
3. Reticulocyte count > 1% (unless Hgb above 10g/dL)
4. No signs significant organ injury (liver/kidney)
5. CXR (if obtained) without lobar infiltrate

C. Past Medical/Social History NEGATIVE for →

Bacteremia/Sepsis
Missing, delayed immunizations (both Prevnar and Pneumovax)
Non-adherence with penicillin prophylaxis
Possibility of non-adherence with follow-up - No phone or Multiple missed appointments or no visit > 1 year in outpatient clinic

*If no phone and no transportation, patient should be admitted for rule out bacteremia

**A working phone number should be obtained directly from the family and transmitted to the Hematology/Oncology Fellow/faculty on call prior to discharge.

*** Pediatric hematology team needs to be notified of all discharges from the ER for fever. The fellow/faculty on call should flag sickle cell team (Kelly Hanlon RN, Emily Braly NP, Arpan Sinha MD, Janna Journeycake MD) regarding all calls so that the team can call and follow up with the patient. Ms. Hanlon, SCD nurse coordinator will follow up with the family within 24 hours. She will notify SW if unable to reach family after 3 attempts in 24 hours.

**** Blood Culture follow up to be done by ER as per their protocol and patient to be called immediately if culture returns positive.

(Updated April 2022 by Arpan Sinha MD, Janna Journeycake MD)