Population: Patients with any sickle hemoglobinopathy (most commonly HbSS, HbSC, HbS Beta\(^0\), or HbS Beta\(^+\)) and Fever > 38.5°C (101°F)

Triage: Should be placed into an examination room as soon as possible.

History and Physical Examination:
- General physical examination for evidence of focal infection
- Evaluation of liver and spleen size (compare to baseline exam in medical records)
- Evaluation of work of breathing and pulse oximetry on room air
- Evaluation of perfusion, degree of pallor
- Neurological examination
- Prior History of Complications: Sepsis, CVA, sequestration, aplastic crisis
- Medications and allergies
- Transfusion history
- It is important to verify immunization history in children with sickle cell, particularly *Hemophilus influenza type b* (Hib) and *pneumococcal* vaccines.
- Ensure that patients less than 6 years old, or those with a history of pneumococcal sepsis, are receiving adequate penicillin prophylaxis:
  - PenVK 125 mg bid (< 3yrs), 250mg bid (> 3yrs)

Diagnostic evaluation:
- CBC with differential, Reticulocyte count
- Blood culture
- Urine culture if indicated (symptomatic, history of UTI, or males under 6mo, girls under 2yrs)
- Consider CXR (chest/upper abd pain, cough, increased WOB, tachypnea, hypoxemia)
- Consider Type and Screen (unusually pale, ill-appearing, or in shock)
  - If a transfusion is required, the blood bank should be notified as cross-matching is more difficult in patients who receiving chronic transfusions (higher Ag-Ab cross-reactivities)
- Attempts should be made to access the medical records. The electronic medical record often contains detailed clinic notes and discharge summaries with a wealth of information (i.e., lab summaries, discharge medications, dosages, prior complications).

Therapeutic interventions:
- Establish IV access
  - D5 NS @ 1-1.5 x maintenance (1200 - 1800cc/m\(^2\)/d)
  - Patients with evidence of decreased perfusion may be bolused with 10-20cc/kg of NS
  - Excess fluids should be avoided
Comer Emergency Department (ED) Clinical Guidelines: Diabetic Ketoacidosis (DKA) – Moderately Severe to Severe

- Administer parenteral antibiotics within 1 hour of presentation to ED (parenteral antibiotics should be given before other procedures – e.g., CXR, etc.)
  - Ceftriaxone 50mg/kg (maximum 2g), or
  - If cephalosporin allergic, Clindamycin 10-15mg/kg/dose (maximum 600mg/dose)
  - Add Azithromycin if infiltrate on CXR (10mg/kg x 1d, 5mg/kg/d x 4)
  - Consider addition of Vancomycin 10-15mg/kg/dose (severely ill, indwelling lines)

- Observe for 1 hour following antibiotics if planning discharge home*
  *Rationale: Acute hemolysis has been described in sickle cell patients receiving antibiotics, most often in association with the administration of ceftriaxone.

- Presence of a focus of infection does not alter the urgency of administration of parenteral antibiotics

- Oxygen recommended if pulse oximetry < 90% on room air, > 5% lower than baseline, acute chest, or other evidence of increased work of breathing (tachypnea, flaring, retractions, tachypnea)

- Antipyretics:
  - Acetaminophen 15 mg/kg/dose q 4hrs
  - Ibuprofen 10mg/kg/dose q 6hrs

Disposition assignment:

- Admission recommended for:
  - Ill-appearing children
  - Age <18 months
  - Prior history of sepsis
  - Fever > 40°C (104°F)
  - WBC > 30k, or < 5k
  - Hgb < 5 g/dL, or > 2g/dL lower than baseline (particularly SC disease)
  - Evidence of poor adherence to recommended therapy or inadequate follow-up (i.e. no hematology visit within 12 months regardless of age).
  - Other associated complications: Suspected aplastic crisis (reticulocyte count < 5% SS, or < 2% SC; or 50% below baseline reticulocyte count)
  - Suspected splenic or hepatic sequestration (enlarging spleen or liver, often with thrombocytopenia or hypotension)
  - Acute chest syndrome (CXR infiltrate ± fever ± oxygen requirement)

Prior to disposition assignment, call the Pediatric Hem/Onc Fellow on call at 4363 for all patients with Sickle Cell Disease and Fever.