DAYTON CHILDREN'S HOSPITAL

CLINICAL PRACTICE GUIDELINES

DISCLAIMER: This Clinical Practice Guideline (CPG) generally describes a recommended course of treatment for patients with the identified health needs. This CPG is not presented and should not be used as a substitute for the advice of a licensed independent practitioner, as individual patients may require different treatments from those specified, and guidelines cannot address the unique needs of each patient. Dayton Children's shall not be liable for direct, indirect, special, incidental or consequential damages related to the use of this CPG.

SICKLE CELL FEVER CLINICAL PRACTICE GUIDELINE NOTES

A. Box 1: Criteria for inclusion

- Genotype HbSS, HbSC, HbS-beta-zero-thalassemia, HbS-beta-plus-thalassemia, HbSD, and all unknown phenotypes, AND
- Temperature > 38.3˚ C (101˚ F)
- Other criteria:
    - Sickle cell patients with painful crises and fever are also included.
    - All sickle cell patients with this degree of fever are included, even if a source of fever (e.g. otitis media) has been identified that is typically treated with oral antibiotics in other individuals.
    - HbAS trait, HbAC trait, and HbCC are excluded from this guideline and can be treated as immune-competent individuals.

B. Box 2: Triage and assessment

- Identification of sickle cell patient with fever should result in immediate triage of the patient so that assessment and subsequent empiric antibiotics can be delivered in a timely fashion (within 60 minutes maximum).
- Assessment: brief history, vital signs and pulse oximetry, fever history, brief physical examination.

C. Box 3: Contact hematologist on call to initiate order set, unless the ED attending physician is immediately available to assess the patient and initiate orders.

D. Box 4: Obtain labs, place PIV, and administer ceftriaxone

- Obtain blood culture, CBC, reticulocyte count.
- Ceftriaxone (Rocephin) 50-100 mg/kg IV should be administered within 60 minutes of presentation to patients who have no known contraindication to this drug.
    - Ceftriaxone (Rocephin) can be given IM if PIV placement is not successful within 45 minutes of arrival in the ED. (Note that the injection may need to be divided into multiple syringes if the maximum volume for an IM injection is exceeded based on patient's weight.)
    - If patient has a contraindication to ceftriaxone, then give ampicillin/sulbactam (Unasyn) 50 mg/kg IV.
    - If patient has a contraindication to ceftriaxone (Rocephin) and PIV placement is not successful within 45 minutes of arrival in the ED, consider ciprofloxacin PO (20 mg/kg).

SICKLE CELL FEVER CLINICAL PRACTICE GUIDELINE NOTES

D. Box 4 (cont.):

- If patient has a contraindication to penicillins and cephalosporins, consult the ED attending physician and consider ciprofloxacin IV (10 mg/kg) or PO (20 mg/kg).

- If blood culture is not obtained within 45 minutes of presentation, administer antibiotic first, and then continue to pursue blood culture.

E. Box 5: Obtain other diagnostic studies as appropriate or relevant, based on the patient's symptoms

- Radiology: chest x-ray (for acute chest syndrome/pneumonia) and other imaging studies as indicated
- Diagnostic lumbar puncture for patients with meningeal signs, neurologic deficit, and/or altered sensorium
- Throat culture
- Stool culture
- Urine culture and urinalysis
- Other

F. Box 6 and 7: Acute chest syndrome?

- Acute chest syndrome should be suspected with any lower respiratory tract symptoms, decreased O_2 saturation, tachypnea, chest pain, cough, or dyspnea.
- If acute chest syndrome is suspected, order pulse oximetry, cardiorespiratory monitoring, incentive spirometry, and oral azithromycin. Blood transfusion may be indicated based on patient's clinical presentation.

G. Box 8: Reassess patient

- Review patient's laboratory results, diagnostic studies (if applicable), and current clinical status.
- **Consult with hematologist on call**

H. Box 9: Ready for discharge?

- Patient may be discharged if <u>all</u> of the following criteria are met:
  - Age greater than 12 months

- Nontoxic, well-appearing

SICKLE CELL FEVER CLINICAL PRACTICE GUIDELINE NOTES

H. Box 9 (cont.):

- Stable vital signs with O_2 sats $\geq$ 92% on room air 60 minutes after antibiotic administration is complete
- Tolerating oral intake
- No concern for complications (i.e., splenic sequestration, acute chest syndrome, or pain crisis requiring IV analgesia)
- Hgb > 5 with no drop greater than 2 g
- WBC > 5,000 and < 30,000
- Chest x-ray (if applicable) without infiltrate
- Patient has reliable follow-up in place (i.e., working telephone, available transportation, no history of missed appointments or noncompliance with medications)

I. Box 10: Patients who meet the discharge criteria should be given a follow-up plan

- For patient with no known source of infection, the discharge physician should contact the hematologist on-call and have the patient follow-up in 24 hours in the Hematology Clinic. (If follow-up is to occur on a day when the Clinic is closed, patient will follow-up in the Almost Home Unit.) **Leave PIV in place**, if possible, for a second dose of ceftriaxone.
- For patient with bacterial source of infection (i.e., Streptococcus, otitis media, urinary tract infection, impetigo), the discharge physician should treat the infection with the appropriate oral antibiotic and have the patient follow-up in the Hematology Clinic after the course of antibiotics is complete.
- For patient with viral source of infection, the discharge physician should contact the hematologist on-call and have the patient follow-up in 24 hours in the Hematology Clinic. (If follow-up is to occur on a day when the Clinic is closed, patient will follow-up in the ED.) **Leave PIV in place**, if possible, for a second dose of ceftriaxone.
- All patients should call their physician or go to the ED for pain, shortness of breath, lethargy, dizziness with standing, or for worsening symptoms.

J. Box 11: Patients who do not meet the discharge criteria should be admitted to the inpatient unit

- Inpatients should have daily CBCs and a daily blood culture for persistent fever.

- Ceftriaxone (or ampicillin/sulbactam) should be continued daily until cultures are negative for a minimum of 48 hours.
- Discharge from inpatient setting when afebrile. Arrange follow-up appointment and consider oral antibiotic.

References

Children's Hospital of Philadelphia. (2015). ED pathway for evaluation/treatment of children with sickle cell disease with fever. Retrieved from http://www.chop.edu/clinical-pathway/sickle-cell-disease-fever-clinical-pathway

Quillen, K., et al. (2008). Prevalence of ceftriaxone-induced red blood cell antibodies in pediatric patients with sickle cell disease and human immunodeficiency virus infection. *The Pediatric Infectious Disease Journal, 27*(4), 357-358.

U.S. Department of Health and Human Services, National Institutes of Health. (2014). Evidence-based management of sickle cell disease expert panel report. Retrieved from http://www.nhlbi.nih.gov/guidelines

Wilimas, J., et al. (1993). A randomized study of outpatient treatment with ceftriaxone for selected febrile children with sickle cell disease. *New England Journal of Medicine, 329*(7), 472-476.