EM CASE OF THE WEEK.

BROWARD HEALTH MEDICAL CENTER
DEPARTMENT OF EMERGENCY MEDICINE

BROWARD HEALTH
Care Warriors

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Acute Sickle Cell Pain Management

A 19-year-old male with a history of sickle cell disease presents to the ED with chest and abdominal pain persisting for the past 4 hours. The pain is described as a throbbing 10/10 pain that is attenuated to a 9/10 after taking Percocet prescribed q6h prn by his pain management specialist. Physical examination is unremarkable. After ruling out life-threatening causes of pain and possible complications of sickle cell disease, which of the following is the most appropriate initial treatment for this patient's pain, according to the NIH Expert Panel on Sickle Cell Disease?

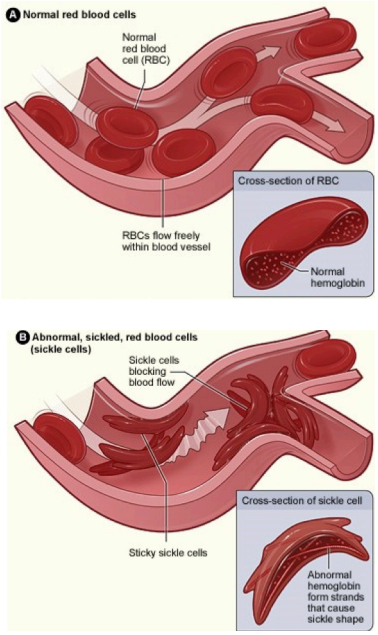
A. Administer Toradol 30 mg IV and reassess in 30 minutes

B. Apply warm compresses to affected area

C. Administer hydroxyurea 15 mg/kg PO

D. Administer Percocet 5/325 mg PO and reassess in 1 hour

E. Administer morphine sulfate 0.1 to 0.15 mg/kg IV and reassess in 30 minutes



National Institute of Health. Sickle Cell Disease. 2017.

(A) shows normal blood flow through vessels (B) shows abnormal blood flow due to sickled red blood cells

EM Case of the Week is a weekly "pop quiz" for ED staff.

The goal is to educate all ED personnel by sharing common pearls and pitfalls involving the care of ED patients. We intend on providing better patient care through better education for our nurses and staff.

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The correct answer is E. According to the National Heart, Lung, and Blood Institute of the NIH Expert Panel on Sickle Cell Disease (SCD), children and adults with SCD in severe vaso-occlusive crisis should be treated within 30 minutes of triage with parenteral opioids such as morphine or hydromorphone (1).

Sickle cell disease is autosomal recessive condition caused by the inheritance of faulty hemoglobin S subunits in place of the typical beta-globin subunits. This leads to the formation of abnormal sickled-shaped red blood cells which have a tendency to aggregate and occlude vasculature causing ischemia, pain, and possible tissue damage. SCD affects approximately 100,000 Americans including 1 out of every 365 African-Americans (2).

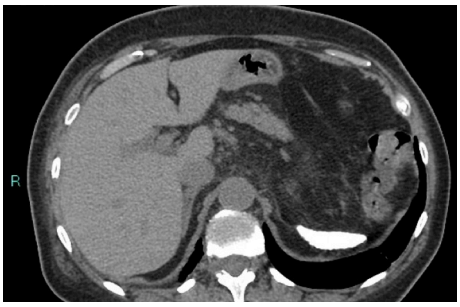
Discussion

Severe pain in sickle cell disease can be representative of vaso-occlusive crisis, the phenomenon caused by the occlusion of microvasculature. Alternatively, it can be representative of tissue infarction and organ damage; therefore, it is very important to rule out potential life-threatening sequelae of SCD in addition to treating the acute episode of pain (Table 1).

Approximately one third of SCD patients experience vaso-occlusive pain by the age of one year old, two thirds by two years old, and ninety percent by six years old (4).

Numerous precipitating factors can lead to vaso-occlusive pain crises. These include dehydration, acidosis, alcohol, cold temperatures, infections, physical exercise, physical or psychologic stress, low oxygen tension, and pregnancy (3).

The most common locations for pain are the upper back, arms, and legs; however, pain in the chest, abdomen, and lower back is also frequently observed. The pain may last for hours to days and may never completely resolve in some patients (4).



Radiopaedia.org. Autosplenectomy. 2017.

Treatment

In an acute setting, the mainstays in treatment in vaso-occlusive crisis are opioids, hydration, and the correction of precipitating factors when possible.

In SCD patients who have previously presented to the ED with similar episodes, prior notes can provide the dosage of opioid utilized as well as the patient's response. This information coupled with the intensity of the pain in the current episode compared to past episodes can serve as a starting point for treatment. In situations where this information is not known, it is suggested to start with either morphine (0.1 to 0.15 mg/kg) or hydromorphone (0.02 to 0.05 mg/kg) and reassess the patient's pain promptly. If the pain is not relieved with more than two dosages, hospitalization for patient controlled analgesia is indicated (4).

In patients who are volume depleted, it is appropriate to replenish the fluid deficit and ongoing losses in order to maintain a euvolemic state; however, there is insufficient evidence to support additional fluid administration (3).

Hydroxyurea should not be routinely begun in the ED as it requires at least three months to demonstrate effectiveness in prevention of vaso- occlusive crises (4).

Finally, whenever possible, any identifiable precipitating factors of the crisis, such as infection, should be promptly corrected.

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ED Work up of Acute Sickle Cell Pain Episode

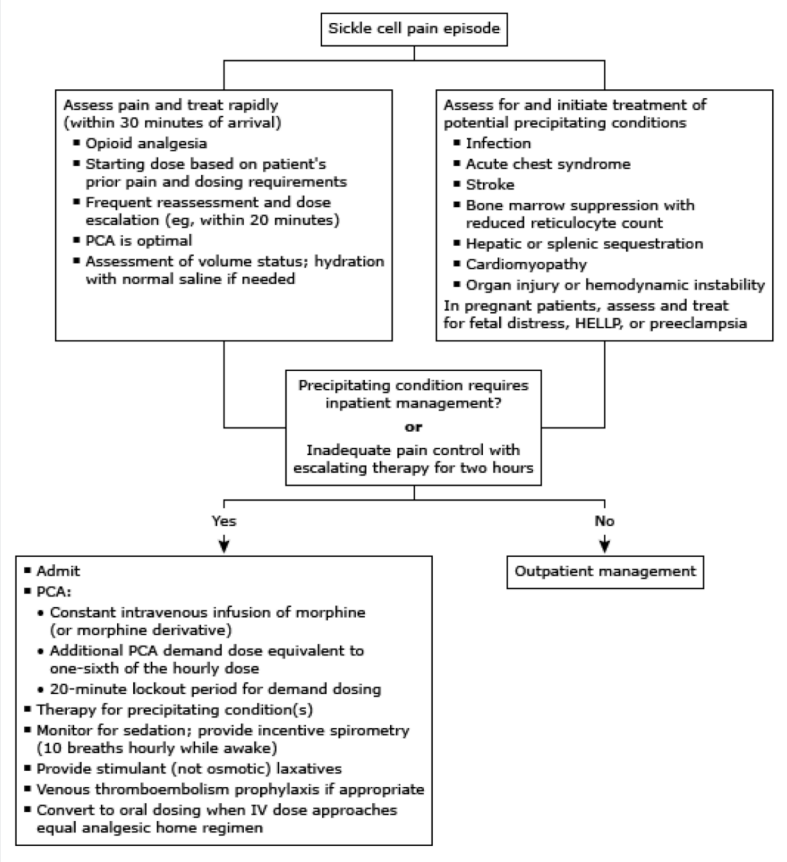

Table 1
UpToDate. Vasoocclusive pain management in sickle cell disease. 2017.

Take Home Points

- Life-threatening causes of pain must be excluded during every ED visit.
- Vaso-occlusive pain episodes are incredibly common in patients with SCD.
- These episodes are organic in nature and should be treated aggressively.
- The choice and dosage of opioid should be guided by the patient's at home regimen, prior successful ED regimens, and the severity of pain in comparison to previous episodes.

ABOUT THE AUTHOR

This month's case was written by Ryan Grell. Ryan is a 4th year medical student from FIU HWCOM. He completed his emergency medicine rotation at BHMC in February 2017. Ryan plans on pursuing a career in Anesthesiology after graduation.

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4. UpToDate. Vasocclusive pain management in sickle cell. 2017 Feb.