EM CASE OF THE WEEK.
BROWARD HEALTH MEDICAL CENTER
DEPARTMENT OF EMERGENCY MEDICINE

BROWARD HEALTH
Care Warriors

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Vol 5 | Issue 38

Acute Pain in Sickle Cell Disease

A 21-year-old female with a history of sickle cell disease presents to the ED with three day onset of worsening 8/10 pain in the fingers, back, and feet with associated shortness of breath at rest, joint pain and stiffness. She has experienced similar symptoms in the past, most recently one month ago, and has been alternating Tylenol #3 and Percocet q4-6 hr without relief. She follows with a hematologist whom she last saw three months ago, and discontinued hydroxyurea four months ago but still takes folate regularly. She denies any fever, nausea/vomiting, chest pain, cough, abdominal pain, or changes in vision. Patient is afebrile, with a BP of 143/81, RR 19, and pulse of 108. On physical exam, patient has enlarged but not obstructing tonsils, and tenderness to palpation of MCPs and PIPs bilaterally without edema, calves, and thoracolumbar spine. Which of the following is the most appropriate initial treatment for this patient's condition?

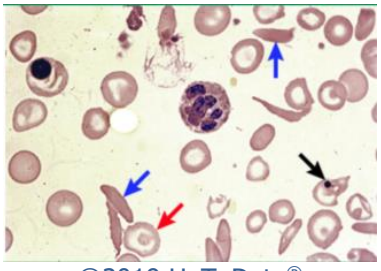
A. Ketorolac

B. Meperidine

C. IV morphine

D. RBC transfusion

E. Cold compress



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Sickled cells (blue arrow) in a blood smear from a patient with SCD

Acute pain is the most common reason patients with SCD come to the emergency department as well as the most frequent complication of SCD.

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 C. IV morphine

Sickle Cell Disease (SCD) is a hemoglobinopathy due to an autosomal recessive mutation that causes a crescent (sickled) shaped deformation of hemoglobin (known as HbS) in red blood cells. This decreases the oxygen carrying capacity of the molecule. The deformed cells are less able to traverse the capillaries and can adhere to vessel endothelium causing inflammation and hallmark symptoms such as vaso-occlusive pain episodes, anemia and organ failure. SCD is the most common inherited blood disorder nationwide, as common as 1/500 African Americans and 1/1000-1400 Hispanics. Symptoms can begin as early as six months old and last throughout the patient's life.[1]

Overview of clinical manifestations of sickle cell disease

	Acute
Pain	Acute vaso-occlusive pain episodes, acute chest syndrome
Infection	Sepsis, pneumonia, meningitis
Anemia	Aplastic crisis, splenic sequestration crisis, hyperhemolytic crisis
CNS	Ischemic stroke, hemorrhagic stroke, seizures, TIA
Pulmonary	Acute chest syndrome, asthma, pulmonary fate embolism, pulmonary thromboembolism
Renal	Renal infarction, medication toxicity, hematuria, acute renal failure, acute nephrotic syndrome
Skeletal	Dactylitis, avascular necrosis
Cardiac	Myocardial infarction, dysrhythmia, sudden death, autonomic dysfunction
Hepatobiliary	Hepatic sequestration crisis, cholecystitis, liver injury, acute intrahepatic cholestasis
Ocular	Retinal artery occlusion, hyphema, retinal detachment
Obstetric	Fetal and maternal complications
Genitourinary	Priapism
Endocrine	
Other	Venous thromboembolism

Discussion

One of the major complications of SCD is vaso-occlusion, which can cause ischemia and infarction of organ tissues as well as hemolytic anemia. There are various etiologies of vaso-occlusive episodes and pain in the acute setting including: acute vaso-occlusive pain; infection; aplastic crisis; acute chest syndrome; renal infarction; dactylitis; splenic sequestration; venous thromboembolism; myocardial infarction; priapism; and avascular necrosis of the hip. Common triggers of acute pain episodes include cold temperature, stress, menstruation, and inadequate hydration, but oftentimes a cause cannot be identified. Multiple areas of pain are often involved and frequently include the long bones and trunk. Acute pain episodes can last up to days and are often followed by an inflammatory stage before resolution. Common lab findings in SCD include Hb between 8-10, Hct 29-30%, mild leukocytosis, and reticulocyte count over 3%.[2,3]

Treatment

It is important to recognize that patients with SCD suffer from both acute and chronic pain, and the goal of treatment in acute pain episodes is to reduce the severity or frequency of pain. Most patients with SCD have debilitating pain, use medications as prescribed and are not seeking drugs.[3]

While multi-organ failure requires immediate treatment with blood transfusion, uncomplicated acute pain episodes do not require exchange therapy. Ideally, patients presenting with acute pain should be assessed rapidly, and analgesia should be given within 30 minutes. Most patients will need IV opioids (morphine or hydromorphone), and dose can be dependent on severity of pain/previous doses or as: morphine 0.1-0.15 mg/kg with a max single dose at 10mg; hydromorphone 0.02-0.05 mg/kg with a max single dose at 1.5mg. Adjunctive medication in addition to (not replacing) IV opioids include IVFs, heat packs, and addressing psychosocial issues. If pain cannot be controlled with three doses, admit to the hospital for patient-controlled-analgesia.[3]

Therapies that should be avoided include cold compresses, meperidine, ketorolac and NSAIDs. This is because cold compresses can induce sickling; meperidine is often needed in multiple doses and can cause CNS toxicity; and the efficacy of ketorolac/NSAIDs have not been proven, and they can cause renal toxicity. Intranasal ketamine in the ED is becoming more popular, however, its recommended use is only in hospitals with a standardized protocol. Supplemental oxygen should only be used if the patient is hypoxic.[3]

Of note, Hydroxyurea and L-glutamine have been shown to reduce the frequency of pain episodes, hospitalizations, and acute chest syndrome, and are a common preventative pharmacotherapy intervention used in SCD.[3]

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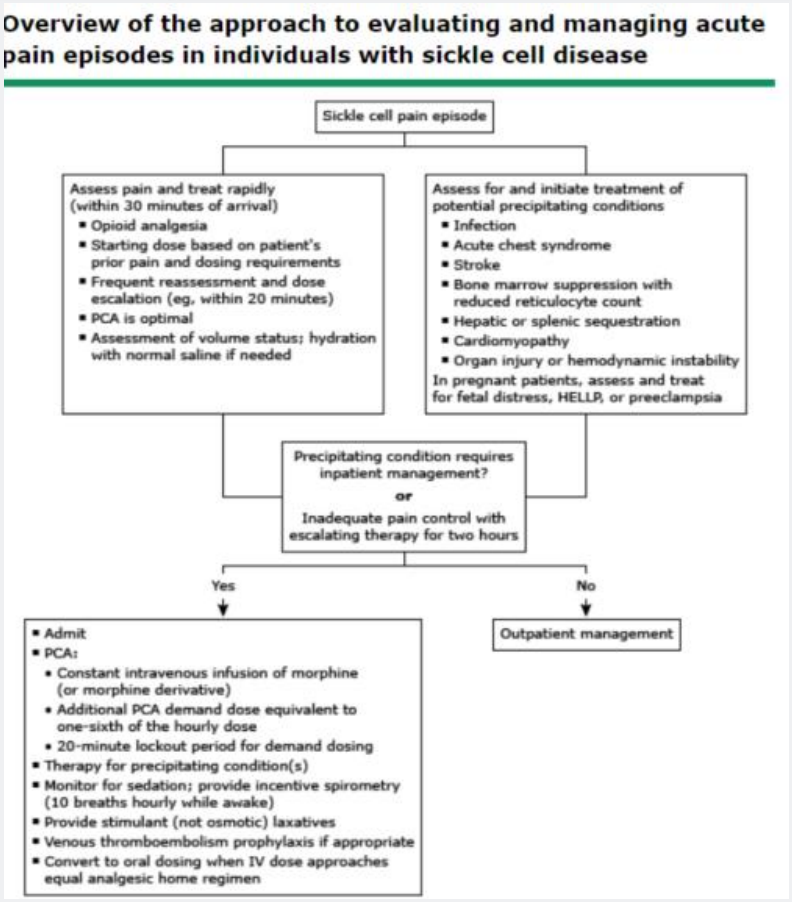

The prognosis of SCD has greatly improved with early detection of disease and infection prevention with vaccines, antibiotics, and medications such as hydroxyurea. An increase in management of SCD complications has also contributed to improved survival and transformation of SCD from a fatal to a chronic process. There are many factors that influence morbidity and mortality with SCD, especially access to care and use of prophylactic antibiotics. In areas where care is accessible, multi-organ failure and acute chest syndrome are the most common causes of death, whereas in areas with more limited resources higher fatalities occur from infections.[4]

Take Home Points

- Vaso-occlusive pain is the most common complication of SCD, often requiring medical attention.
- Acute pain may be isolated, or may be related to an underlying, potentially fatal complication of SCD. Thus, patients should be thoroughly and rapidly evaluated for life-threatening etiologies while treating pain.
- The gold standard for evaluation of pain is patient self-report.
- While chronic pain may be managed at home by many patients with SCD, the mainstay treatment for acute pain in the emergent setting is IV opioid analgesics. Avoid ketorolac and meperidine.

This case was written by Kristina Chung. Kristina is a 4th year medical student from FIU. She did her emergency medicine rotation at BHMC in January 2019. Kristina plans on pursuing a career in Pediatrics after graduation.

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