CASE 10.2
Sickle Cell Disease | Level 2

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1. What subjective and objective evidence supports the diagnosis of a sickle cell pain crisis and of acute chest syndrome?

PAIN CRISIS

SUBJECTIVE FINDINGS: Self-reported pain started as 5/10 on VAS and increased to 10/10; pain described as sharp, throbbing, worse with movement and with deep breaths; home pain medications did not help decrease pain; in moderate distress

OBJECTIVE FINDINGS: Increased pulse, increased respiratory rate

ACUTE CHEST SYNDROME

SUBJECTIVE FINDINGS: Described chest pain on left side, worse with deep breaths; reported nonproductive cough for 4 days

OBJECTIVE FINDINGS: CXR shows no new infiltrates, but infiltrates may take a day or two to manifest; chest auscultation demonstrates decreased aeration on left; decreased oxygenation per pulse oximeter reading; increased respiratory rate; increased temperature

2. Develop a pharmacologic regimen for the treatment of the pain crisis and the suspected acute chest syndrome in this patient.

Patients with pain that cannot be adequately controlled on home medications, as well as any patient with SCD who has chest pain or signs and symptoms of acute chest syndrome, should be admitted for inpatient therapy. Goals of therapy for the treatment of pain crisis are to relieve pain (at least back to their baseline), increase hemoglobin levels if decreased, maintain adequate hydration, and prevent complications from the use of opioids (e.g., itching, nausea, sedation, respiratory depression, and constipation). It is unrealistic to expect patients to have no pain; because this is a chronic disease some level of pain is almost always present, but pain should be controlled as much as possible for patients to be able to function well and adequately perform activities of daily living.

Goals of therapy for acute chest syndrome include maintaining oxygen saturations at or above 92% on room air; treating bacterial infections if present (or at least treat empirically until resolution of symptoms of infection such as fever, increased WBC count, chest pain, and cough); resolving symptoms such as chest pain and increased respiratory rate;

increasing hemoglobin levels if decreased; and maintaining adequate hydration.

PAIN CRISIS

The patient should receive IV fluids at 1 to 1.5 times maintenance for a total of 2,380 mL/day or 100 mL/hr. She also has suspected acute chest syndrome, so IV fluids should be limited to maintenance rates to avoid fluid overload. Because of a hemoglobin level of 6.5 g/dL, she should be transfused with pRBCs 640 mL (10 mL/kg) over 4 hours. All patients with SCD who require transfusions should be transfused with leukocyte-poor, antigen-matched blood to reduce the frequency of transfusion reactions and the development of antibodies. Generally, transfusion should only be administered if the hemoglobin level is 1.5 times below baseline or less than 7 g/dL, and the patient requires oxygen supplementation.

The severe pain the patient is experiencing, self-rated as 10/10 and not responding to oral pain medications, is being treated with a morphine PCA with a dose of 1.3 mg (0.02 mg/kg) IV every 10 minutes with a basal of 3.8 mg (0.02 to 0.06 mg/kg/hr). The use of the higher basal dosing is appropriate given her uncontrolled pain and pain ratings of 10/10. Alternately, she could receive a continuous infusion of morphine at 2 mg/hr (0.02 to 0.05 mg/kg/hr) titrated to effect, or intermittent IV dosing of 5 mg (0.05 to 0.15 mg/kg) IV every 3 to 4 hours prn. In this patient's case, the use of the PCA with basal dosing will give her more control over her intermittent dosing and may result in an overall lower opioid dose administered, which may decrease adverse reactions from the morphine such as constipation, itching, drowsiness, respiratory depression, and sedation. She should also receive an NSAID, such as ketorolac 30 mg (0.5 mg/kg) IV every 6 hours scheduled for at least 48 hours and up to 5 days. Ketorolac should not be used for longer than 5 continuous days due to increased incidence of adverse reactions such as gastrointestinal bleeding. After 5 days of ketorolac, she may be changed to ibuprofen 600 mg (10 mg/kg rounded to the nearest tablet size) po every 6 to 8 hours until the pain crisis has resolved.

The morphine therapy may cause constipation; therefore, a stool softener should be initiated prophylactically. Docusate 100 mg po tid (5 mg/kg/day divided in one to four doses) was ordered at admission. Consider changing to a laxative, such as polyethylene glycol 3350 (MiraLAX®) 17 g mixed in water or juice once daily, if the patient complains of constipation. Opioid adverse reactions also include itching, which may be treated initially with diphenhydramine 25 mg (1 mg/kg/dose) IV or po every 6 hours (max 25 to 50 mg/dose) and if itching remains uncontrolled a low-dose naloxone drip of 16 mcg/hr IV infusion (initiate at 0.25 and increase to no more than 1 mcg/kg/hr if necessary), concomitant with the morphine PCA or continuous infusion should be considered. If she develops nausea or vomiting from the use of the opioid, ondansetron 4 mg (0.1 mg/kg/dose with max dose of 4 mg) IV or po every 8 hours, or promethazine 16 mg (0.25 mg/kg) IV or po every 6 hours for a maximal dose of 25 mg/dose, may be useful. Ondansetron should be initiated first to prevent nausea and vomiting; it generally has fewer adverse reactions such as sedation, than promethazine; however, once the patient has started vomiting, the promethazine should be started because ondansetron is not as effective for treating vomiting as promethazine. If the adverse reactions of the morphine become unmanageable, it may be helpful to change to a different opioid such as hydromorphone.

ACUTE CHEST SYNDROME

Fluid therapy should be monitored closely; while necessary, it is important to avoid fluid overload, which may lead to pulmonary edema and increased respiratory distress. In light of this patient having both acute chest syndrome suspected as well as a known pain crisis, IV fluids should be kept at no more than daily maintenance requirements. Close attention should be paid to her daily intake and output to ensure fluid balance. Oxygen per nasal cannula should be started to keep the oxygen saturation at greater than 92%. Because her hemoglobin is low (6.2 g/dL), she should receive a transfusion of 640 mL (10 mL/kg) of pRBCs. Her hemoglobin should be monitored, and a goal of her normal baseline hemoglobin should be made; the goal is to get the patient back to her base-