



Nursing Practice Guidelines: Care of the Patient with Sickle Cell Disease at Risk for/With Acute Chest Syndrome

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Introduction

The International Association of Sickle Cell Nurses and Physician Assistants has developed this guideline with the intention of promoting interaction among disciplines to ensure consistency of practice for patients with sickle cell disease with or at risk for acute chest syndrome (ACS). This nursing practice guideline applies to any inpatient or outpatient at risk for or with ACS and is not intended to limit or dictate practice for the treatment of ACS.

ACS is defined as an acute illness characterized by new pulmonary infiltrate on the chest x-ray and some degree of chest pain, dyspnea, fever and hypoxemia. It should be noted that changes in the chest x-ray often lag behind the clinical presentation. Acute chest syndrome is a frequent cause of death in both children and adults and can occur in any type of sickle cell disease. The lungs are the site where deoxygenated sickle cells pass through capillaries to pick up oxygen; they are at an increased risk for vasoocclusion. Risk factors include: high baseline leukocyte count, bed rest/immobility, use of opioids, over-hydration, and low baseline hemoglobin F percent. Causes of acute chest include: infection, infarction, vascular damage and inflammation, atelectasis, hypoventilation, and bronchospasm.

Assessment

The nurse should assess every patient for risk of acute chest syndrome on admission and regularly throughout the hospitalization.

The nurse should have a high degree of suspicion for acute chest syndrome when any of the following are present:

- Prior history of ACS/pneumonia
- Acute or progressive respiratory changes (use of accessory muscles, nasal flaring, shallow respirations, adventitious breath sounds, decreased breath sounds, shortness of breath, dyspnea, pain, or cough)
- Fever
- Chest pain/abdominal pain
- Oxygen saturation below normal for patient
- Recent general anesthesia



Other risk factors from the medical history include:

- Prior acute chest syndrome/pneumonia (number of episodes, date of last episode, need for transfusions or intubation)
- Asthma, reactive airway disease, or other pulmonary disorder
- Cardiovascular disease
- Recent general anesthesia
- Smoking or exposure to second-hand smoke
- Recent history of prolonged bed rest (at home) or inactivity
- Increased use of prescribed opiate analgesics
- Pain inconsistent with the patient's usual pain crises

Elements of the hospitalization itself that can increase the risk for acute chest syndrome:

- Prolonged bed rest and inactivity
- Poor respiratory effort
- Administration of high dose opiates and the resulting sedation
- Over hydration with IVF and or blood products

***Note that patient can present with none of these symptoms and develop ACS within a few hours.**

Intervention

- Assess the patient at least every 2 hours for the following: vital signs, oxygen saturation, comprehensive respiratory status, mental status changes, cardiovascular status, strict I&O.
- Complete blood count, reticulocyte count, blood cultures, type and cross match, chest x-ray as clinically indicated.
- Provide analgesics for adequate pain control
- Broad spectrum antibiotics and coverage for atypical organisms
- Encourage use of incentive spirometer
- Bronchodilators as indicated
- Supplemental oxygen to treat hypoxemia.
- Intubation and mechanical ventilation may be necessary if impending respiratory failure is suspected.
- Transfusion therapy may be indicated in order to optimize oxygen carrying capacity and to interrupt the cycle of hypoxemia and sickling. This can be done as a simple transfusion or as exchange transfusion. See transfusion guidelines and your hospital transfusion protocol.

Documentation

- Documentation should be done according to institutional requirements, but should include vital signs, medications, treatments, patient response, pain assessment, education and discharge planning

Education

- Adequate treatment of underlying cardiovascular or pulmonary pathology. Assess compliance with prescribed treatments
- Patients should live in a smoke-free environment. Refer to a smoking cessation program as indicated
- Frequent use of incentive spirometry and avoidance of over hydration and prolonged bed rest
- Necessity of completing prescribed course of antibiotics
- Importance of ambulation
- Seek medical attention for fever greater than or equal to 101°F, chest pain or difficulty breathing
- Importance of recommended medical follow- up
- Consider initiation of Hydroxyurea therapy (see *Nursing Practice Guideline for Care of the Patient with Sickle Cell Disease Receiving Hydroxyurea*)
- Consider the use of short term transfusion therapy (see *Nursing Practice Guideline for Care of the Patient with Sickle Cell Disease Receiving Transfusion Therapy*)

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