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Common Acute Complications of Sickle Cell Disease (SCD)

Complication	Risk Factors and Presentation	Evaluation	Transfusion Indicated?	Management
Acute chest syndrome (ACS)	ACS resembles pneumonia, eg, the patient has signs and symptoms of lower respiratory tract disease and a new pulmonary infiltrate on chest radiograph. ACS can develop suddenly or insidiously. Patients may present with fever, tachypnea, chest pain, and hemoptysis. Risk factors include recent infection, surgery, and dehydration.	Chest radiograph, CBC, and arterial blood gas analysis.	Transfusion is indicated for severe ACS.	Supportive care, including oxygen, hydration, and pain management. Transfusion is the mainstay of treatment.

Administration of Opioids for Severe, Acute Pain

Use an individualized prescribing and monitoring protocol
(*C e , E e Pa e*).

Do not use meperidine unless it is the only effective opioid for
an individual patient (*C e , Ada ed*).

Calculate the parenteral (IV or subcutaneous) dose based on
total daily short-acting opioid dose currently being taken at home
(*C e , E e Pa e*).

Administer parenteral opioids using the subcutaneous route
when intravenous access is difficult (*C e , E e Pa e*).

Reassess pain and re-administer opioids if necessary for
continued severe pain every 15–30 minutes until pain is under
control per patient report (*C e , Ada ed*).

Maintain or consider escalation of the dose by 25 percent until
pain is controlled (*C e , E e Pa e*).

Reassess after each dose for pain relief and side effects
(*C e , E e Pa e*).

Initiate around-the-clock opioid administration by patient-
controlled analgesia or frequently scheduled doses versus "as
requested" administration (*M de a e ec _ _ e da , , ' -
q a , e , de ce*).

In individuals with a vaso-occlusive crisis (VOC) who require
antihistamines for itching secondary to opioid administration,
prescribe agents orally, and do not re-administer with each dose

This pocket guide is adapted from the National Heart, Lung, and Blood Institute's *Evidence-Based Management of Sickle Cell Disease: Expert Panel Report*, 2014, available at <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4061442/>. Two companion pocket guides adapted from the same report are available: "Health Maintenance and Management of Chronic Complications of Sickle Cell Disease" and "Hydroxyurea and Transfusion Therapy for the Treatment of Sickle Cell Disease."

This guide is not intended to be construed as a standard of care or to preempt clinical judgment. Recommendations based on expert opinion or less than high-quality evidence should inform shared decisionmaking with the patient about diagnostic and treatment alternatives. Even recommendations based on high-quality evidence may be inappropriate for some patients depending on clinical circumstances including individual patient preferences.

Dr. Desai is a consultant for Pfizer. Dr. McCavit is a consultant for Pfizer and was previously a consultant for GlycoMimetics.

To order this and other pocket guides, go to www.ama-assn.org/spe.

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