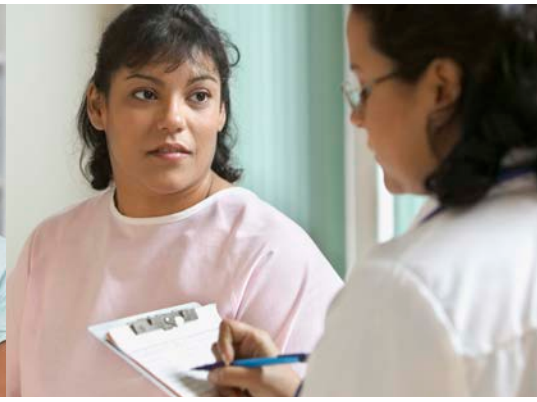
Evidence-Based Management of Sickle Cell Disease

Expert Panel Report, 2014: Guide to Recommendations





U.S. Department of Health and Human Services
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2. In consultation with a sickle cell expert, transfuse people who have acute splenic sequestration and severe anemia to raise the hemoglobin to a stable level, while avoiding over-transfusion.
(Strong Recommendation, Low Quality Evidence)

3. In consultation with a sickle cell expert, address the performance and timing of splenectomy in people with recurrent acute splenic sequestration or symptomatic hypersplenism.
(Moderate Recommendation, Low-Quality Evidence)

Acute Chest Syndrome

1. Evaluate people with SCD who develop acute onset of lower respiratory tract disease signs and/or symptoms (cough, shortness of breath, tachypnea, retractions, or wheezing) with or without fever for ACS. This should include a chest x ray and measurement of oxygen saturation by pulse oximetry.
(Consensus–Panel Expertise)

2. Hospitalize people with ACS.
(Consensus–Panel Expertise)

3. Treat people with SCD who have ACS with an intravenous cephalosporin, an oral macrolide antibiotic, supplemental oxygen (to maintain oxygen saturation of greater than 95 percent), and close monitoring for bronchospasm, acute anemia, and hypoxemia.
(Strong Recommendation, Low-Quality Evidence)

4. In people with SCA, give simple blood transfusion (10 mL/kg red blood cells) to improve oxygen carrying capacity to people with symptomatic ACS whose hemoglobin concentration is >1.0 g/dL below baseline. If baseline hemoglobin is 9 g/dL or higher, simple blood transfusion may not be required.
(Weak Recommendation, Low-Quality Evidence)

5. In people with HbSC disease or HbSβ+-thalassemia with ACS, decisions about transfusion should be made in consultation with an SCD expert.
(Strong Recommendation, Low-Quality Evidence)

6. In all persons with SCD, perform urgent exchange transfusion—with consultation from hematology, critical care, and/or apheresis specialists—when there is rapid progression of ACS as manifested by oxygen saturation below 90 percent despite supplemental oxygen, increasing respiratory distress, progressive pulmonary infiltrates, and/or decline in hemoglobin concentration despite simple transfusion.
(Strong Recommendation, Low-Quality Evidence)

7. Encourage use of incentive spirometry while awake.
(Strong Recommendation, Moderate-Quality Evidence)

Acute Stroke

1. In people with SCD who present with severe headache, altered level of consciousness, seizures, speech problems, and/or paralysis, evaluate for acute stroke by seeking neurologic consultation and performing an urgent head computerized tomography (CT) scan followed by magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) if available.
(Consensus–Panel Expertise)

2. In consultation with a sickle cell expert, perform exchange transfusion in people with SCD who develop acute stroke confirmed by neuroimaging.
(Consensus–Panel Expertise)

3. Initiate prompt evaluation, including neurologic consultation and neuroimaging studies, in people with SCD who have mild, subtle, or recent history of signs or symptoms consistent with transient ischemic attack.
(Consensus–Panel Expertise)