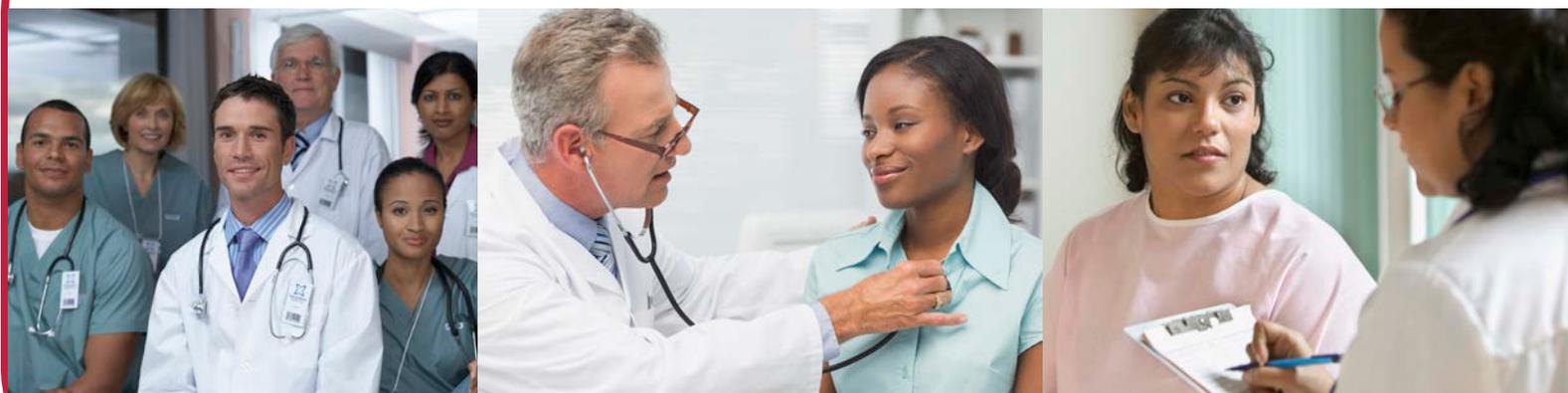


Evidence-Based Management of Sickle Cell Disease

Expert Panel Report, 2014: Guide to Recommendations



U.S. Department of Health and Human Services
National Institutes of Health
National Heart, Lung, and Blood Institute

www.nhlbi.nih.gov

Chapter 3: Managing Acute Complications of Sickle Cell Disease

Acute Anemia

1. During all acute illnesses in people with SCD, obtain a CBC and reticulocyte count, repeat daily in all hospitalized patients, and compare the results with the patient's prior measurements.
(Consensus–Panel Expertise)
2. Assess people with SCD whose hemoglobin concentration is 2 g/dL or more below their baseline (or less than 6 g/dL when the baseline is unknown) for acute splenic sequestration, an aplastic episode, a delayed hemolytic transfusion reaction, ACS, and infection.
(Consensus–Panel Expertise)
3. Use simple transfusion in people with SCD and acute anemia whose symptoms are due to anemia.
(Consensus–Panel Expertise)
4. Perform a CBC and reticulocyte count promptly and again 7 to 10 days later in siblings and others with SCD who are exposed to a person with an aplastic episode.
(Consensus–Panel Expertise)
5. Manage aplastic events with immediate red blood cell transfusion aimed at restoring the hemoglobin to a safe (not necessarily baseline) value. Isolation of hospitalized patients (droplet precautions) is required to prevent spread of the parvovirus B19 to pregnant women and others with SCD or compromised immunity.
(Consensus–Panel Expertise)

Splenic Sequestration

1. In people with hypovolemia due to severe acute splenic sequestration, immediately provide IV fluid resuscitation.
(Strong Recommendation, Low-Quality Evidence)

Chapter 3: Managing Acute Complications of Sickle Cell Disease

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)