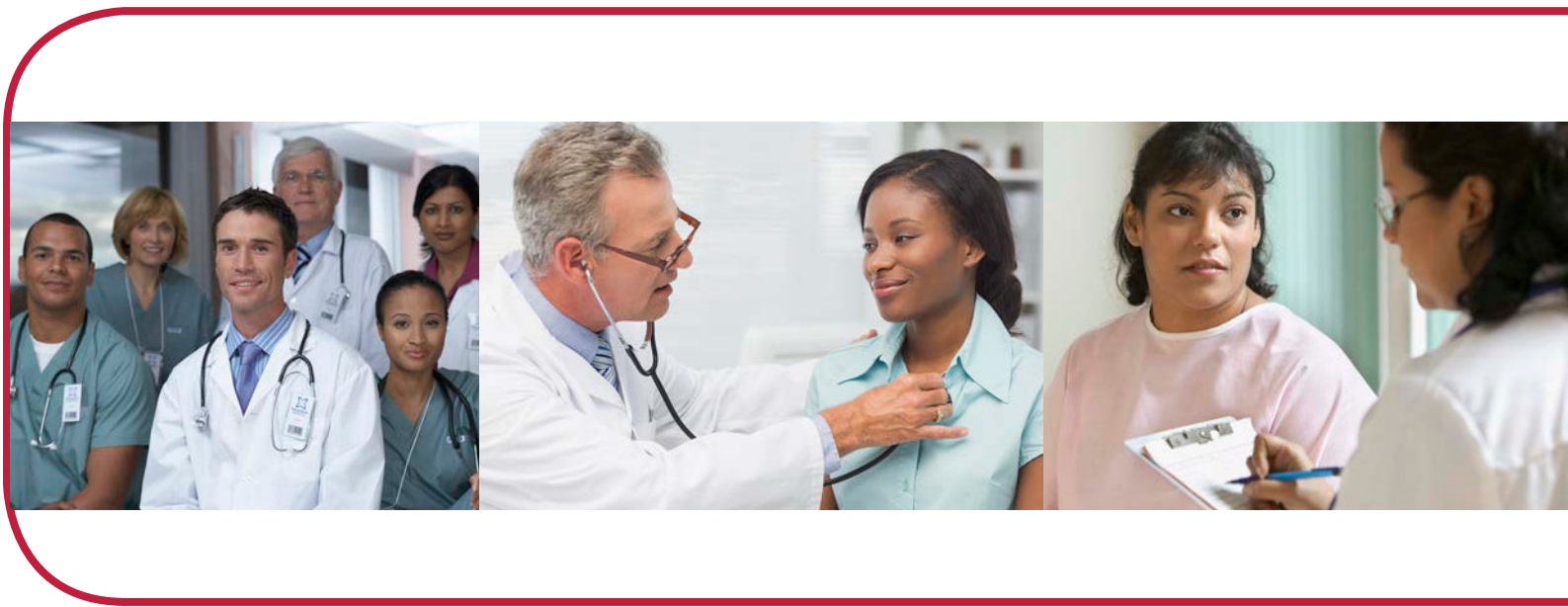


# Evidence-Based Management of Sickle Cell Disease

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](http://www.nhlbi.nih.gov)

## Chapter 3: Managing Acute Complications of Sickle Cell Disease

5. In people with HbSC disease or HbS $\beta$ -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)**

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4. In children and adults who have had a stroke, initiate a program of monthly simple or exchange transfusions.  
**(Moderate Strength, Low-Quality Evidence)**
5. In children and adults who have had a stroke, if it is not possible to implement a transfusion program, initiate hydroxyurea therapy.  
**(Moderate Strength, Low-Quality Evidence)**

### Multisystem Organ Failure

1. In people with SCD who exhibit severe deterioration during a VOC, immediately evaluate for potential MSOF.  
**(Consensus–Panel Expertise)**
2. In people with SCD and respiratory failure, support respiratory status with supplemental oxygenation and mechanical ventilation when needed.  
**(Consensus–Panel Expertise)**
3. Use renal replacement therapy (e.g., hemodialysis) when needed for acute renal failure.  
**(Consensus–Panel Expertise)**
4. In people with SCD and MSOF, immediately initiate either simple or exchange transfusion in consultation with a sickle cell expert or hematologist.  
**(Consensus–Panel Expertise)**