



# Effective Health Care

## Prevention of Sickle Cell Disease Complications in Adults Nomination Summary Document

### Results of Topic Selection Process & Next Steps

- The topic, *Prevention of Sickle Cell Disease Complications in Adults*, was addressed by a 2014 National Heart, Lung, and Blood Institute (NHLBI) guideline, which provides comprehensive evidence-based recommendations for preventive care and management of acute and chronic complications in adult sickle cell disease (SCD) patients.
  - National Heart, Lung, and Blood Institute (NHLBI). *Evidence-based management of sickle cell disease: Expert panel report, 2014*. Bethesda (MD): National Heart, Lung, and Blood Institute (NHLBI), 2014. Accessed October 8, 2015. Available at: <https://www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines>

### Topic Description

**Nominator(s):** Individual  
**Nomination Summary:** People with sickle cell disease (SCD) are living longer yet the long-term health complications associated with SCD in adulthood have not been well studied. As a result, the nominator is concerned that the adult population is being left behind when it comes to preventive healthcare, leading to premature disability and death. The nominator asserts that the early death rate is too high and this disparity is related to complications of SCD that are occurring in adulthood. The nominator is interested in discovering if early interventions that address SCD complications will result in prolonged patient health & reduced health care cost. The nominator intends to pass this information to the legislative and medical partners of his community. According to the nominator, the results could impact health care cost and thereby cause changes in treatment protocols.

**Staff-Generated PICO Population(s):**

**Population:** Adults(18 years and older) with SCD

**Intervention(s):** Interventions to prevent complications, including use of hydroxyurea, routine transfusions, immunization, and other preventive measures

**Comparator(s):** Other interventions to prevent and complications, standard care, or no care

**Outcome(s):** Acute SCD complications (e.g., stroke, acute chest syndrome); chronic SCD complications (e.g., hemosiderosis, chronic pain, pulmonary hypertension); morbidity; mortality

**Key Questions from Nominator:** For adult patients with sickle cell disease what are the comparative related health complications which lead to morbidity? As a result, are there preventative measures that could be taken?

We contacted the nominator to ask for further clarification on the topic nomination. Based on the additional information provided by the nominator, and in consultation with our clinical reviewer, we further refined the Key Question as follows:

In adults with sickle cell disease, what are effective measures for preventing and managing complications of sickle cell disease that occur during adulthood?

## Considerations

- The topic meets EHC Program importance criteria. (For more information, see <http://effectivehealthcare.ahrq.gov/index.cfm/submit-a-suggestion-for-research/how-are-research-topics-chosen/>.)
- The life expectancy of a person with sickle cell disease (SCD) is much longer than in the past. This has led to the need for interventions to prevent and manage SCD complications and improve health outcomes.
- Topic was found to be addressed by a 2014 National Heart, Lung, and Blood Institute (NHLBI) guideline, which provides comprehensive evidence-based recommendations for preventive care and management of acute and chronic complications in adult with SCD. The guideline provides recommendations for the general health maintenance of individuals with SCD; prevention of acute SCD complications (i.e., vaso-occlusive crises, fever, acute renal failure, priapism, hepatobiliary complications, acute anemia, splenic sequestration, acute chest syndrome, acute stroke, multisystem organ failure, acute ocular conditions); and prevention of chronic SCD complications (i.e., chronic pain, avascular necrosis, leg ulcers, pulmonary hypertension, renal complications, stuttering/recurrent priapism, and ophthalmologic complications).