

New guideline created for managing sickle cell disease

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An expert panel has created a new evidence-based guideline for managing sickle cell disease (SCD), with a strong recommendation for the use of the drug hydroxyurea and transfusion therapy for many individuals with SCD, although high-quality evidence is limited, with few randomized clinical trials conducted for this disease, according to an article in the September 10 issue of *JAMA*.

Sickle cell disease is a life-threatening genetic disorder affecting nearly 100,000 individuals in the United States; most of those affected are of African ancestry or self-identify as black. This disease is associated with many acute and chronic complications requiring immediate medical attention. Care for persons with SCD often lacks continuity. Primary care and emergency care health professionals need up-to-date clinical guidance regarding care of persons with this disease, according to background information in the article.

In 2009, the National Heart, Lung, and Blood Institute convened an [expert panel](#) that developed the Evidence-Based Management of Sickle Cell Disease: Expert Panel Report 2014. Barbara P. Yawn, M.D., M.Sc., M.S.P.H., of the Olmsted Medical Center, Rochester, Minn., and colleagues produced a summary of this report and conducted a search of the medical literature to examine the strength of the quality of evidence for the recommendations.

Among the strong recommendations:

- For prevention: daily oral prophylactic penicillin up to the age of 5 years, annual transcranial Doppler examinations from the ages of 2 to 16 years in those with [sickle cell anemia](#), and long-term [transfusion therapy](#) to prevent stroke in those children with abnormal transcranial Doppler velocity (elevated speed of blood flow in the cerebral arteries).
- To address acute complications: rapid initiation of opioids for treatment of severe pain associated with a vasoocclusive crisis (blockage of blood flow due to the abnormal "sickled" red blood cells getting stuck in the blood vessels) and use of incentive spirometry (a method of encouraging deep breathing with the use of an instrument to provide feedback) in patients hospitalized for a vasoocclusive crisis.
- For chronic complications: use of analgesics and physical therapy for treatment of avascular necrosis (a condition in which poor blood supply to an area of bone leads to bone death) and use of angiotensin-converting enzyme inhibitor therapy for microalbuminuria (a subtle increase in the urinary excretion of the protein albumin that cannot be detected by a conventional urinalysis) in adults with SCD.
- For children and adults with proliferative sickle cell retinopathy: referral to expert specialists for consideration of laser photocoagulation and for echocardiography to evaluate signs of pulmonary hypertension.

A recommendation of moderate strength suggests offering treatment with hydroxyurea without regard to the presence of symptoms for infants, children, and adolescents.

The authors note that some of the strong recommendations appearing in this guideline are supported by low- or very low-quality evidence, and that evidence is lacking in many areas important to care for individuals with SCD.

"The process of developing guidelines for the management of children, adolescents, and adults with SCD has been challenging because high-quality evidence is limited in virtually every area related to SCD management. The systematic review of the literature identified a very small number of [randomized clinical trials](#) in individuals with SCD, demonstrating the extensive knowledge gaps in SCD and care of affected individuals. The expert panel realizes that this summary report and the guidelines leave many uncertainties for health professionals caring for individuals with SCD and highlight the importance of collaboration between primary care health professionals and SCD experts. However, we hope that this summary report and the SCD guideline begins to facilitate improved and more accessible care for all affected individuals, and that the discrepancies in the existing data will trigger new research programs and processes to facilitate future guidelines," the authors conclude.

Michael R. DeBaun, M.D., M.P.H., of the Vanderbilt University School of Medicine, Nashville, Tenn., comments on this guideline in an accompanying editorial.

"Yawn and colleagues have undertaken a monumental effort to produce a practical, evidence-based guideline for SCD. Many aspects of this guideline will help both individuals with the disease and clinicians. As would be expected, when the guideline is based on recommendations from randomized clinical trials, such as penicillin prophylaxis, transcranial Doppler screening, blood transfusion therapy prior to surgery, or hydroxyurea therapy for severe disease, these strong recommendations will be embraced by the SCD community. However, when recommendations are based on consensus panel expertise, practice variation will justifiably continue."

More information: Paper: [DOI: 10.1001/jama.2014.10517](https://doi.org/10.1001/jama.2014.10517)
Editorial: [DOI: 10.1001/jama.2014.11103](https://doi.org/10.1001/jama.2014.11103)

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