A large share of children with sickle cell anemia are not receiving recommended care to prevent dangerous complications of the disease—specifically, screening to prevent strokes or recommended medication to stave off other disease-related problems.

This finding, from a recent report from the Centers for Disease Control and Prevention (CDC), “highlights the disheartening fact that many young people are not receiving potentially life-saving screenings and treatment for this disease,” said Laura A. Schieve, PhD, of the CDC’s National Center on Birth Defects and Developmental Disabilities and the report’s lead author, at a press briefing. The report also underscores how surveillance efforts by the CDC and participating states can provide information that points to the need to establish new sickle cell disease specialty clinics in areas where they are currently lacking, guide new research initiatives, and inform the development of state health care policies.

Sickle cell disease (a group of blood cell disorders that includes its most severe form, sickle cell anemia) is one of the most common inherited blood disorders in the United States, but the exact number of people living with it is unknown. The disease primarily affects persons with African ancestry, and its complications include anemia; pain; infections; pneumonia; a severe lung-related complication called acute chest syndrome; stroke; and kidney, liver, and heart disease. The disease is also associated with a reduced life expectancy.

In 2014, an expert panel issued guidelines to prevent or reduce complications of sickle cell disease, several of which affect youth in particular. In children and adolescents aged 2 years to 16 years with sickle cell anemia, the panel recommended annual screening with transcranial Doppler ultrasound to identify those at risk for stroke, and advised hydroxyurea therapy for individuals 9 months or older to reduce the risk for several life-threatening complications.

For those children and teens identified as being at risk for stroke, blood transfusions can substantially lower the likelihood of stroke. In addition, research has demonstrated that hydroxyurea is effective in preventing or reducing severe pain episodes and other complications of sickle cell anemia and in increasing survival.

In the new report, the researchers examined Medicaid records for 3352 children and adolescents (aged 2 years to 16 years) with sickle cell anemia to see if they had received ultrasound screening, hydroxyurea, or both. Even though ultrasound screening had increased 27% among individuals aged 10 years to 16 years during 2014 to 2019, fewer than half (47%) of children aged 2 years to 9 years and only 38% of children and teens aged 10 years to 16 years received ultrasound screening in 2019.

During the same period, hydroxyurea use also increased significantly among children younger than 10 years (27%) and children and adolescents aged 10 years to 16 years (23%). Still, in 2019, only 38% of children aged 2 years to 9 years and slightly over half (53%) of children and adolescents aged 10 years to 16 years used hydroxyurea.

The CDC says that a number of barriers contribute to less-than-optimal care for patients with sickle cell disease, such as a shortage of clinicians with expertise in treating patients with the disorder and with familiarity with guidelines for ultrasound screening and hydroxyurea use. Logistic issues, such as dealing with regular clinician and laboratory visits for monitoring hydroxyurea therapy, limited availability of appointments for transcranial Doppler ultrasound screening, or the
need to travel long distances to radiology centers that perform such screening, also can be problematic.

The report also notes that given that more than 90% of patients with sickle cell disease are Black and up to 9% are Hispanic, racism and health care disparities compound barriers to care for children with sickle cell anemia. Prejudice, discrimination, and bias toward patients with sickle cell anemia may lead to inadequate care, and policies that have led to unequal opportunities in housing, employment, health insurance, and research funding “keep disparities in place and contribute to adverse health outcomes,” the investigators wrote.

Preventing complications associated with sickle cell anemia requires strategies to reduce racism and disparities, the report says, noting that clinicians “can educate themselves, their colleagues, and their institutions about the unique and specific needs of persons with [sickle cell anemia], including how racism impedes optimal health care.”

To address gaps in care, more comprehensive data on various populations with sickle cell disease, such as that collected by the Sickle Cell Data Collection (SCDC) program, are needed. This program, a state-based tracking system established by the CDC in 2015 in California and Georgia, analyzes data from participating states that highlight health care needs and gaps to guide new research initiatives and the development of state health care policies.

For example, as detailed in another report released last month by the CDC, which included SCDC data for Georgia and California, geographic assessments informed decision-making on the need for additional community resources and sickle cell disease specialty care clinics in these states. In Georgia, the analysis revealed that 10% of infants born with sickle cell disease during 2004 to 2016 lived more than a 1-hour drive from the closest option for specialty care, leading to the opening of new mobile care clinics. In California, public health officials found that in 2016 to 2018, most of the 1800 patients with the disorder in Los Angeles County lived approximately 15 to 60 miles from hematologists with experience in treating patients with sickle cell disease—highlighting a need to establish more specialty care clinics.

By 2021, the SCDC program expanded to include 11 states (Alabama, California, Colorado, Georgia, Indiana, Michigan, Minnesota, North Carolina, Tennessee, Virginia, and Wisconsin), which collectively cover approximately 36% of persons with sickle cell disease in the US. As additional funding becomes available, the SCDC program will expand to new states, which will allow researchers to obtain a more complete picture of people with sickle cell disease throughout the US.

For individuals living with sickle cell anemia and sickle cell disease, such an expansion of surveillance coverage “would allow CDC to better characterize disease outcomes and health care needs … across the life span,” Schieve and her colleagues wrote.

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**ARTICLE INFORMATION**

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