September 14, 2022

The Honorable Xavier Becerra  
Secretary  
Department of Health and Human Services  
200 Independence Avenue, SW  
Washington, DC 20201

The Honorable Chiquita Brooks-LaSure  
Administrator  
Centers for Medicare and Medicaid Services  
7500 Security Boulevard  
Baltimore, MD 21244

Robert Otto Valdez, PhD, MHSA  
Director  
Agency for Healthcare Research and Quality  
5600 Fishers Lane  
Rockville, MD 20857

Dear Secretary Becerra, Administrator Brooks-LaSure, and Director Valdez:

The 17 undersigned organizations commend the Department of Health and Human Services (HHS), the Centers for Medicare and Medicaid Services (CMS), and the Agency for Healthcare Research and Quality (AHRQ) on the commitment to improve health equity and reduce health disparities. As an extension of your efforts to date, we strongly urge HHS to adopt the following National Quality Forum (NQF) measures for inclusion in the Medicaid Child Core Measure Set:

- NQF measure #2797: Transcranial Doppler Ultrasonography Screening Among Children with Sickle Cell Anemia  
  - Assesses the percentage of children ages 2 through 15 years of age with sickle cell anemia (SCA, i.e., Hemoglobin SS and Hemoglobin Sβthalassemia) who received at least one transcranial Doppler (TCD) ultrasonography screening within the measurement year.

- NQF measure #3595: Hydroxyurea Use Among Children with Sickle Cell Anemia  
  - Assesses the percentage of children with SCD who were dispensed hydroxyurea for at least 300 days within the measurement year.

The most common cause of stroke in children is sickle cell disease (SCD). Without screening and prophylactic treatment, between 5 and 17% of individuals with SCD will suffer a first stroke during childhood or adolescence. This is also an important health equity issue. Stroke among children with SCD, most of whom are Black, is largely preventable with proper screening and treatment, yet a significant percentage of these
children do not receive the recommended care. TCD ultrasonography is a safe, non-invasive, low-cost screening technique that measures blood velocities within the cerebral vessels. It has been shown to accurately identify children at higher risk for stroke, allowing preventive treatment to be initiated.

Hydroxyurea and chronic blood transfusions are the only currently proven disease-modifying treatments for children with SCD. Both therapies are used in primary and secondary stroke prevention. Although neither has been shown to prevent all SCD-related strokes, these treatment modalities can significantly reduce the risk of stroke and improve the quality of life for individuals with SCD. Further, hydroxyurea has been shown to significantly reduce the incidence of pain episodes and acute chest syndrome. Treatment with hydroxyurea is underutilized for many people with SCD who could benefit from it.

The National Heart, Lung, and Blood Institute (NHLBI), the American Society of Hematology (ASH), and the American Stroke Association division of the American Heart Association (AHA) all recommend that all eligible children with SCD receive TCD screening annually from age 2 through 16 years. They also recommend that children who have abnormal TCD screening results who cannot receive regular blood transfusion therapy (due to availability, affordability, or family preference) should be offered hydroxyurea treatment at the maximum tolerated dose to substitute for regular blood transfusions. In addition, in their 2016 report to CMS and again in 2018, the Measures Applications Partnership recommended the addition of NQF #2797 to strengthen the Medicaid Child Core Set and address this high-priority gap area. These measures are feasible for states to collect, since they use administrative claims data and do not require costly and time-intensive chart review. Both measures have been thoroughly tested and received consensus endorsement from NQF in 2016 and 2021, respectively, but have not been adopted by Medicaid.

Additionally, the American Society of Hematology 2020 guidelines recommend additional strategies to identify children with SCD and silent cerebral infarcts who may be at risk for cognitive impairment. Please see Attachment 1 for more information about these recommendations. Together, the clinical application of these recommendations and the TCD and hydroxyurea measures will significantly decrease the neurological burden of SCD in children at greatest risk for poor academic attainment and decrease the health equity gap between Black and White students. We encourage HHS, CMS, and other partners to consider these recommendations when considering the development of additional measures for SCD.

In closing, despite their importance in primary and secondary prevention of strokes and recommendation in national guidelines, TCD ultrasonography screening and hydroxyurea remain significantly underused. Fewer than 4 out of 10 (36 percent) of Medicaid and Children’s Health Insurance Program (CHIP) beneficiaries ages 2 through 15 with SCD had at least one TCD screening and only 16 percent of eligible Medicaid and CHIP beneficiaries ages 21 months to 20 years, received hydroxyurea for at least half the year in 2017. This substantial gap in quality of care disproportionately affects children.
who are Black in the United States. Adoption of these measures as Medicaid Child Core Measures would contribute to closing this significant quality gap. Implementation of these measures would help avert serious adverse events for children with SCD as well as support the ongoing efforts of HHS, CMS, and AHRQ to advance health equity. We hope these measures can be considered for inclusion in the Medicaid Child Core Measure Set at the earliest opportunity.

Thank you for your time and consideration. If you have any questions or need any additional information, please contact Emily Holubowich, American Heart Association Vice President, Federal Advocacy, at emily.holubowich@heart.org or 202.785.7912.

Sincerely,

AABB
American Academy of Neurology
American Heart Association
American Society of Hematology
American Society of Pediatric Hematology/Oncology
Association of Pediatric Hematology/Oncology Nurses
Association of Public Health Laboratories
Children’s Healthcare of Atlanta
Foundation for Sickle Cell Disease Research
International Alliance for Pediatric Stroke
MUSC Shawn Jenkins Children’s Hospital
Pediatric Hospital Sickle Cell Disease Collaborative
Sick Cells
Sickle Cell 101
Sickle Cell Consortium
Sickle Cell Disease Association of America, Inc. (SCDAA)
Texas Children’s Hospital

Cc:
Dr. Lee Fleisher, CMS Chief Medical Officer and Director, Center for Clinical Standards and Quality
Daniel Tsai, Deputy Administrator, and Director CMCS
Dr. Aditi Mallick, CMO, CMCS
Dr. Kamila Mistry, Director Division of Priority Populations Research, Senior Advisor for the Child Health and Quality Improvement
Kimberly Miller-Tolbert, MPH, Health Policy Advisor, Office of the Secretary, HHS
Dr. Michelle S. Davis, Senior Advisor for Public Health Strategy, Office of the Assistant Secretary for Health, HHS
Attachment 1

Additional Recommendations re: Evaluating Neurological Complications in Sickle Cell Disease (SCD)

As highlighted in the cover letter, our groups encourage the Department of Health and Human Services (HHS) to adopt the National Quality Forum (NQF) measure for Transcranial Doppler Ultrasonography Screening Among Children with Sickle Cell Anemia for inclusion in the Medicaid Child Core Measure Set. Additionally, since the NHLBI guidelines were published, there is now better understanding of other neurological complications in sickle cell anemia (SCA) and other types of SCD. Approximately 35% of all children with Hemoglobin SS and Hemoglobin Sβ0 thalassemia, less than 18 years of age will have a silent cerebral infarct (SCI). SCIs are not overt strokes with obvious neurological impairments, but abnormalities seen on MRI imaging. Importantly they are associated with a five-point decrease in their Full-Scale IQ score when compared to children with SCD without SCIs. Furthermore, there is a high prevalence of developmental delay and cognitive impairments in children with all types of SCD. Based on this, the American Society of Hematology 2020 guidelines recommended the following strategies to identify children with SCD and SCIs as well as any child with SCD who may be at risk for cognitive impairment. We encourage HHS, CMS and/or other SCD stakeholders to consider the following recommendations when developing new measures for SCD.

- **Recommendation 9.1.** For children with SCD and abnormal screening for developmental or cognitive status, the ASH guideline panel recommends the following:
  - A developmental, cognitive, and medical evaluation to diagnose any related disorders and to identify modifiable risk factors for developmental delays or cognitive impairments.
  - Following the cognitive domain-specific evidence-based guidelines for these conditions to provide appropriate interventions.
  (Strong recommendation based on high certainty in the evidence about effects ⬤⬤⬤⬤).

- **Recommendation 10.1.** Given the high prevalence of SCIs in children with SCD (1 in 3), and their association with cognitive impairment, poor school performance, and future cerebral infarcts, the ASH guideline panel recommends:
  - At least a 1-time MRI screening, without sedation, to detect SCIs in early-school-age children, when MRI can commonly be performed without sedation.
  (Strong recommendation based on moderate certainty in the evidence about effects ⬤⬤⬤⬤).  

Thus, we encourage HHS to adopt NQF measure #2797 as well as encourage HHS, CMS, or other SCD stakeholders to consider the additional ASH guideline recommendations when developing new measures for SCD aimed at preventing and detecting neurological injury. Together, these three recommendations will significantly decrease the
neurological burden of SCD in children at greatest risk for poor academic attainment and decrease the health equity gap between Black and White students.