

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 S β ⁰thalassemia) 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.¹ 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.² 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.³

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.^{2, 4, 5} 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.^{6, 7} 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 2017.⁸ 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 S β^0 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.

¹ Kirkham, F.J.; Lagunju, I.A. Epidemiology of Stroke in Sickle Cell Disease. *J. Clin. Med.* 2021, 10, 4232. Available at: <https://doi.org/10.3390/jcm10184232>.

² Kolata, Gina. "These Sisters With Sickle Cell Had Devastating, and Preventable, Strokes." *New York Times*, May 23, 2021. <https://www.nytimes.com/2021/05/23/health/sickle-cell-black-children.html>.

³ Evidence-Based Management of Sickle Cell Disease Expert Panel Report, 2014. Available at: [Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014 | NHLBI, NIH](#).

⁴ DeBaun MR, Jordan LC, King AA, et al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults Available at: <https://doi.org/10.1182/bloodadvances.2019001142>.

⁵ Meschia JF, Bushnell C, Boden-Albala B, Braun LT, Bravata DM, Chaturvedi S, Creager MA, Eckel RH, Elkind MSV, Fornage M, Goldstein LB, Greenberg SM, Horvath SE, Iadecola C, Jauch EC, Moore WS, Wilson JA; on behalf of the American Heart Association Stroke Council, Council on Cardiovascular and Stroke Nursing, Council on Clinical Cardiology, Council on Functional Genomics and Translational Biology, and Council on Hypertension. Guidelines for the primary prevention of stroke: a statement for healthcare professionals from the American Heart Association/American Stroke Association. *Stroke*. 2014;45:3754–3832.

⁶ Strengthening the Core Set of Healthcare Quality Measures for Children Enrolled in Medicaid and CHIP, 2016.

https://www.qualityforum.org/Publications/2016/08/Strengthening_the_Core_Set_of_Healthcare_Quality_Measures_for_Children_Enrolled_in_Medicaid,_2016.aspx.

⁷ Strengthening the Core Set of Healthcare Quality Measures for Children Enrolled in Medicaid and CHIP, 2018.

https://www.qualityforum.org/Publications/2018/08/Strengthening_the_Core_Set_of_Healthcare_Quality_Measures_for_Children_Enrolled_in_Medicaid_and_CHIP,_2018.aspx.

⁸ Wilson-Frederick S, Hulihan M, Mangum A, Khan T, Geibel M, Malsberger R, Verghese S, Borck R, Fox R, Rosenbach M. Medicaid and CHIP Sickle Cell Disease Report, T-MSIS Analytic Files (TAF) 2017. Baltimore, MD: Center for Medicaid and CHIP Services, Division of Quality and Health Outcomes, Centers for Medicare & Medicaid Services, 2021.