blood advances

TO THE EDITOR:

Hematologist encounters among Medicaid patients who have sickle cell disease

Sophia S. Horiuchi,[1] Mei Zhou,[2] Angela Snyder,[2] and Susan T. Paulukonis[1]

[1]Tracking California Program, Public Health Institute, Richmond, CA; and [2]Georgia Health Policy Center and the Department of Public Management and Policy, Andrew Young School of Policy Studies, Georgia State University, Atlanta, GA

Sickle cell disease (SCD) is a group of inherited hemoglobin disorders, affecting individuals from birth throughout their lifespan. Although life expectancy for those with SCD has increased into adulthood in recent decades,[1] the effects of the disease are cumulative and lead to significant illness and reduced quality of life for many.[2] Hallmarks of this disease include acute and chronic pain episodes, risk of organ damage and stroke, and high acute health care utilization.[3] SCD affects ~100 000 persons in the United States, and millions more globally.[4] In the United States, because SCD affects primarily Black persons and other persons of color, there are increased challenges for patients, families, and providers owing to institutional and systemic racism.[5,6]

Access to comprehensive, quality care for those with SCD has been shown to improve outcomes and lower acute care utilization (hospitalization and emergency room encounters), particularly among adults.[7-10] Although comprehensive care for children with SCD is not universally available, there are centers providing such care in most US urban areas.[11] Health care providers specializing in nonmalignant hematology for adults, however, are rare.[12,13] Upon transitioning out of pediatric care, adults encounter a dearth of providers knowledgeable in SCD and a fragmented health care system.[13] Multiple models have been proposed for best practice care for SCD,[14-17] but even the simplest of these models, that of a hematologist and primary care physician comanaging a patient, is infrequently seen in practice.[17]

Although various barriers to care have been identified,[15] the percentage of the SCD population that do not receive care is unknown. The Sickle Cell Data Collection (SCDC) program is a state-based, population-wide public health surveillance system for SCD. SCDC data from 2 states with large populations of people living with SCD, California and Georgia, were analyzed to quantify how often those with SCD receive care from a hematologist.

A retrospective analysis was performed on Medicaid claims for individuals with SCD. Individuals with SCD are identified in each state by linking, de-duplicating, and applying a validated case definition to multisourced surveillance data that span from 2004 through 2019.[18-20] Individuals from the states' SCDC cohorts with continuous Medicaid enrollment from January 1, 2016 to December 31, 2018 were included in this analysis to capture all inpatient and outpatient health care encounters. Patient age was calculated for the first day of the study period and categorized as pediatric (<21 years) or adult (21+ years).

Hematologist encounters were identified using the National Provider Identifier of the rendering provider listed in claims.[21] Providers with health care provider taxonomy codes 207RH0000X, 207RH0003X, or 2080P0207X were categorized as a hematologist. To calculate an individual's total hematologist encounters, claims were de-duplicated by assuming an individual had a maximum of 1 encounter with a given provider per day.

Submitted 22 March 2022; accepted 4 July 2022; prepublished online on *Blood Advances* First Edition 12 July 2022; final version published online 1 September 2022. DOI 10.1182/bloodadvances.2022007622.

Individual data cannot be shared (even deidentified) by data use agreement with the states of California and Georgia. Aggregate data are available at https://www.cdc.gov/ncbddd/hemoglobinopathies/scdc-data.html.

All analyses were performed using SAS software, version 9.4. The SCDC programs and this study were overseen by review of the California Committee for the Protection of Human Subjects and the Georgia State University institutional review board under an exemption for public health surveillance.

Among individuals with SCD that were enrolled in Medicaid at any time in 2016 to 2018, 79% in California and 52% in Georgia had continuous enrollment for the entire 3-year period and therefore met the inclusion criteria for this analysis. There were 989 pediatric patients and 1919 adults in California, and 1711 pediatric patients and 1373 adults in Georgia (Table 1). Georgia had a greater percentage of pediatric patients (55%) than California (35%). States had a similar sex distribution (California: 41% male; Georgia: 45% male).

The proportions of the pediatric patients in our samples who had *no encounters with a hematologist* in the 3-year period were 24% in California and 13% in Georgia (Figure 1). Among adults included in our study samples, 56% in California and 34% in Georgia had *no encounters with a hematologist* in the 3-year period. The median number of hematologist encounters in the 3-year period for the pediatric population was 7 in California and 8 in Georgia, whereas for adults, the median was 0 in California and 3 in Georgia (Table 1).

Among patients who had a hematologist encounter in the 3-year period, 99% of the pediatric patients in both states were seen at least once in an outpatient setting by a hematologist, and among adults, 90% in California and 91% in Georgia.

One gap in the literature is data on how many of those living with SCD are seen with regularity by a hematologist knowledgeable in SCD care. This limitation is due to the lack of a national surveillance system that can identify individuals who are not connected to care and thus missing from registries and clinical cohorts. In addition, there is no agreement on how to define a provider knowledgeable in SCD care.[22] The National Heart, Lung, and Blood Institute's SCD Implementation Consortium has defined sickle cell specialists with 9 criteria related to training and experience.[23] However, data on these criteria are not available. Given this limitation, the SCDC programs investigated whether persons with SCD had seen any hematologist, the expected primary specialist for a hemoglobinopathy. In California and Georgia, 24% and 13% of pediatric patients covered by Medicaid from 2016 to 2018 had no encounters with a hematologist in a 3-year period. For adults, the proportion was significantly higher, 56% for California and 34% for Georgia.

Table 1. Description of individuals with SCD continuously enrolled in Medicaid in California and Georgia, 2016-2018

	California	Georgia
Total (n)	2908	3084
Median age, y	29.6 (IQR = 25.5)	17.0 (IQR = 24)
Pediatric, n (%)	989 (34)	1711 (55)
Median pediatric age, y	12.0 (IQR = 8)	9.0 (IQR = 8)
Median pediatric hematologist encounters in 3-y period	7 (IQR = 24)	8 (IQR = 15)
Adult, n (%)	1919 (66)	1373 (45)
Median adult age, y	38.7 (IQR = 20)	34.0 (IQR = 19)
Median adult hematologist encounters in 3-y period	0 (IQR = 4)	3 (IQR = 15)
Sex, n (%)		
Male	1177 (41)	1393 (45)
Female	1731 (59)	1691 (55)

IQR, interquartile range.

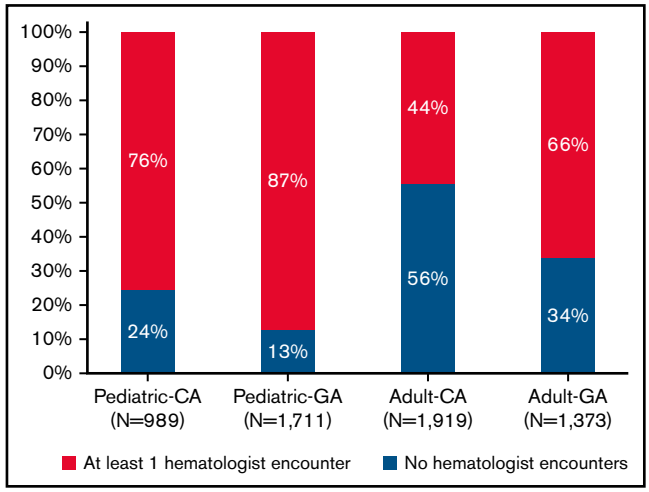

Figure 1. For individuals with SCD and continuous Medicaid coverage from 2016 to 2018, all encounters with a hematologist were identified using Medicaid claims. Individuals were categorized into those that had no hematologist encounters or those that had at least one hematologist encounter during the 3-year period from 2016 to 2018.

In contrast to SCD, chronic genetic diseases such as cystic fibrosis and hemophilia have networks of comprehensive specialty care.[14] A recent study found that 82% of persons with hemophilia and 94% with severe hemophilia received care at a federally funded comprehensive specialty center in a 3-year period.[24] Although the population with SCD in the United States is over double that of hemophilia, there is no equivalent system of care for SCD. There is an opportunity for this patient population to be brought into quality care settings to effectively manage their disease and prevent complications and mortality.

Not all hematologists have the expertise and experience necessary to provide care for a disease as complex as SCD. There were 765 and 342 hematologists in California and Georgia, respectively, who saw 1 or more patients meeting the inclusion criteria for this study. Of these hematologists, 66% saw <5 such patients in California and 40% in Georgia during the 3-year period. Whether these hematologists had training or background knowledge in SCD care is unknown, but the high proportion with few patients suggests that centralization of care is lacking.

Hematologists are not the only knowledgeable SCD providers; primary care providers (PCPs) may offer preventative, therapeutic, and pain care for adults with the disease and may even specialize in the care of this population. We also investigated whether there could be such providers and their contribution to care among these populations. Providers with a primary health care provider taxonomy code for nurse practitioners, physician assistants, or medical doctors with specialties listed as Internal Medicine, Pediatrics, Family Medicine, or General Practice were categorized as a PCP. A PCP

experienced in SCD care was defined as a PCP with 20 or more patients with SCD in the 3-year period that met the inclusion criteria for this analysis. For pediatric patients that had no encounters with a hematologist in the 3-year period, we found that 10% in California and 36% in Georgia were seen by these experienced PCPs in the same period. For adults, these percentages were 18% in California and 37% in Georgia. Care by an experienced and well-trained non–hematologic care provider can be the primary source of SCD care for those with the disease, but this group of providers is only partially filling the gap in SCD care in these 2 states. All told, 46% of California adults with SCD and 21% of those in Georgia still had no encounters with a hematologist or an experienced PCP over the 3-year period.

This analysis was limited to Medicaid claims data. Health care utilization for individuals that were privately insured, uninsured, or covered by Medicare, or Medicaid patients that did not have continuous enrollment during the study period, is not captured in these results. However, Medicaid is a major provider of insurance for the SCD population. Over a 3-year period, 66% of people with SCD in California and 56% in Georgia were insured by Medicaid.[25] These surveillance data provide a unique look into access to care issues.

Individuals living with SCD in 2 states with high SCD populations are seeing hematologists at a lower rate than in populations with other chronic genetic diseases, particularly adults. More research and investigation into the individuals who are not seen by SCD experts, and the reasons for this lack of access to appropriate care, is critical to understand workforce development, infrastructure, outreach, and other needs to address this complex chronic disease.

Acknowledgments: The authors thank Mary Hulihan of the Division of Blood Disorders, Centers for Disease Control and Prevention (CDC) for her thoughtful review and comments on this manuscript.

This study is supported by funding from grant CDC-RFA-DD20-2003 and was previously supported by the CDC Foundation, the Doris Duke Charitable Foundation, Pfizer Inc, Global Blood Therapeutics, and Sanofi Inc.

The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the CDC.

Contribution: S.S.H. designed and conducted the analysis, wrote the manuscript; M.Z. contributed to the design of the surveillance system and provided technical assistance in data interpretation, conducted analysis for Georgia data; A.S. contributed to the design of the surveillance system and provided technical assistance in results interpretation; and S.T.P. contributed to the design of the surveillance system and design of the study.

Conflict-of-interest disclosure: The authors declare no competing financial interests.

ORCID profile: S.T.P., 0000-0002-0663-7657.

Correspondence: Susan T. Paulukonis, Tracking California, 31 Branham St NE, Atlanta, GA 30317; e-mail: susan.paulukonis@trackingcalifornia.org.

References

1. Quinn CT, Rogers ZR, McCavit TL, Buchanan GR. Improved survival of children and adolescents with sickle cell disease. *Blood.* 2010; 115(17):3447-3452.
2. Blinder MA, Vekeman F, Sasane M, Trahey A, Paley C, Duh MS. Age-related treatment patterns in sickle cell disease patients and the associated sickle cell complications and healthcare costs. *Pediatr Blood Cancer.* 2013;60(5):828-835.
3. CDC. What is sickle cell disease? Available at: https://www.cdc.gov/ncbddd/sicklecell/facts.html. Accessed 1 December 2020.
4. Brousseau DC, Panepinto JA, Nimmer M, Hoffmann RG. The number of people with sickle-cell disease in the United States: national and state estimates. *Am J Hematol.* 2010;85(1):77-78.
5. Power-Hays A, McGann PT. When actions speak louder than words – racism and sickle cell disease. *N Engl J Med.* 2020; 383(20):1902-1903.
6. Lee L, Smith-Whitley K, Banks S, Puckrein G. Reducing health care disparities in sickle cell disease: a review. *Public Health Rep.* 2019; 134(6):599-607.
7. Lanzkron S, Carroll CP, Hill P, David M, Paul N, Haywood C Jr. Impact of a dedicated infusion clinic for acute management of adults with sickle cell pain crisis. *Am J Hematol.* 2015;90(5): 376-380.
8. Basishvili G, Gotesman J, Vandervoort K, Jacobs C, Vattappally L, Minniti CP. Comprehensive management reduces incidence and mortality of acute chest syndrome in patients with sickle cell disease. *Am J Hematol.* 2018;93(3):E64-E67.
9. Koch KL, Karafin MS, Simpson P, Field JJ. Intensive management of high-utilizing adults with sickle cell disease lowers admissions. *Am J Hematol.* 2015;90(3):215-219.
10. Yang YM, Shah AK, Watson M, Mankad VN. Comparison of costs to the health sector of comprehensive and episodic health care for sickle cell disease patients. *Public Health Rep.* 1995;110(1):80-86.
11. CDC. Sickle Cell Disease National Resource Directory. Available at: https://stacks.cdc.gov/view/cdc/11903. Accessed 1 December 2020.
12. Sobota A, Neufeld EJ, Sprinz P, Heeney MM. Transition from pediatric to adult care for sickle cell disease: results of a survey of pediatric providers. *Am J Hematol.* 2011;86(6):512-515.
13. Kanter J, Smith WR, Desai PC, et al. Building access to care in adult sickle cell disease: defining models of care, essential components, and economic aspects. *Blood Adv.* 2020;4(16): 3804-3813.
14. Grosse SD, Schechter MS, Kulkarni R, Lloyd-Puryear MA, Strickland B, Trevathan E. Models of comprehensive multidisciplinary care for individuals in the United States with genetic disorders. *Pediatrics.* 2009;123(1):407-412.
15. NASEM (National Academies of Sciences, Engineering, and Medicine). Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. Washington, DC: The National Academies Press; 2020.
16. Okpala I, Thomas V, Westerdale N, et al. The comprehensiveness care of sickle cell disease. *Eur J Haematol.* 2002;68(3):157-162.
17. Crego N, Douglas C, Bonnabeau E, et al. Sickle-cell disease co-management, health care utilization, and hydroxyurea use. *J Am Board Fam Med.* 2020;33(1):91-105.
18. Hulihan MM, Feuchtbaum L, Jordan L, et al. State-based surveillance for selected hemoglobinopathies. *Genet Med.* 2015;17(2):125-130.

19. Paulukonis ST, Harris WT, Coates TD, et al. Population based surveillance in sickle cell disease: methods, findings and implications from the California registry and surveillance system in hemoglobinopathies project (RuSH). *Pediatr Blood Cancer.* 2014; 61(12):2271-2276.

20. Snyder AB, Zhou M, Theodore R, Quarmyne MO, Eckman J, Lane PA. Improving an administrative case definition for longitudinal surveillance of sickle cell disease. *Public Health Rep.* 2019;134(3):274-281.

21. Bindman AB. Using the National Provider Identifier for health care workforce evaluation. *Medicare Medicaid Res Rev.* 2013;3(3): mmrr.003.03.b03.

22. Kanter J, Meier ER, Hankins JS, Paulukonis ST, Snyder AB. Improving outcomes for patients with sickle cell disease in the United States: making the case for more resources, surveillance, and longitudinal data. *JAMA Health Forum.* 2021;2(10):e213467.

23. Sickle Cell Disease Implementation Consortium. Unaffiliated patients. Available at: https://scdic.rti.org/OUR-PROGRESS/Unaffiliated-Patients. Accessed 22 October 2021.

24. Okolo AI, Soucie JM, Grosse SD, et al. Population-based surveillance of haemophilia and patient outcomes in Indiana using multiple data sources. *Haemophilia.* 2019;25(3): 456-462.

25. Center for Disease Control. Medicaid coverage patterns for people with sickle cell disease (SCD) in California and Georgia, 2014-2016. Available at: https://www.cdc.gov/ncbddd/hemoglobinopathies/documents/scdc-fact-sheet-medicaid-data-h.pdf. Accessed 21 October 2021.