SICKLE CELL DISEASE TREATMENT DEMONSTRATION REGIONAL COLLABORATIVES PROGRAM

Report to Congress

SEPTEMBER 2021

NICHQ
National Institute for Children's Health Quality

HRSA
Maternal & Child Health

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Executive Summary

Ensuring that the approximately 100,000 children and adults living with sickle cell disease (SCD) and their families are offered and obtain current lifesaving treatments – and that they have equitable access to quality care and successful new treatments and cures for this condition – stands as a national priority.

Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program

In 2017, through a Health Resources and Services Administration (HRSA), Department of Health and Human Services Funding Opportunity Announcement, five Regional Coordinating Centers (RCCs) were chosen to establish a Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP). This program was originally authorized by the American Jobs Creation Act of 2004, Title VII, § 712(c), P.L. 108-357, Title VII, § 712(c)(42 U.S.C. 300b) (2004) and was reauthorized by the Sickle Cell Disease and Other Heritable Blood Disorders Research, Surveillance, Prevention, and Treatment Act of 2018, 42 U.S.C. § 300b-5 (2018). The Program was administered and funded by the HRSA Maternal and Child Health Bureau (MCHB), Division for Children with Special Health Care Needs.

Built from the progress of the prior SCD programs reported to Congress in 2014 and 2017, the purpose of the funding was to support RCCs in establishing regional networks and enable them to provide leadership and support for regional and state activities that would develop and establish system-wide mechanisms to improve the prevention and treatment of SCD and collect select data in these three healthcare domains:

1. Increase the number of providers treating individuals with SCD using the National Heart, Lung and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report (National Heart Lung and Blood Institute, 2014)

NOTE: Throughout this report, the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program is referred to as "the Program" or "the SCDTDRCP."

2. Use telementoring, telemedicine, and other provider strategies to increase the number of providers administering evidence-based SCD care
3. Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

The goals were to improve health outcomes in individuals with SCD, reduce morbidity and mortality caused by SCD, reduce the number of individuals with SCD receiving care only in emergency departments, and improve the quality of coordinated and comprehensive services to individuals with SCD and their families.

Overview of Sickle Cell Disease

Approximately 100,000 Americans live with SCD, which refers to a group of inherited red blood cell disorders whereby red blood cells become hard and sticky, die early, and tend to impede blood flow leading to serious health problems. While this disease affects many races and ethnicities, in the U.S. it disproportionately affects Black, African-American, and Hispanic-American populations (Centers for Disease Control and Prevention, 2020). One of every 400-500 Black and about one out of every 16,300 Hispanic-American babies are born with SCD each year (Benson & Therrell Jr, 2010; Centers for Disease Control and Prevention, 2020; Feuchtbaum et al., 2012; Kato et al., 2018) making it one of the most common serious genetic disorders in the U.S. (Neumayr et al., 2019; Rees et al., 2010). Although historically SCD was considered a pediatric condition because children with SCD did not survive into adulthood, with medical advances and improvements in care most people living with SCD now survive into adulthood. In the mid-1970s, people with SCD lived to a median age of 14. By 2017, the median age had increased to 43 years. This represents a little more than half the national average lifespan of 81 years for women and 77 for men. Despite these gains, people living with SCD face a lifelong battle with pain, infection, and other chronic, serious health problems that can affect every organ in the body.

Core Program Partners

1. Five Regional Coordinating Centers (RCC)

The RCCs established a regional infrastructure and network comprised of clinical locations and community-based organizations (CBOs) covering the U.S. states and territories. Each region had at least one designated regional lead. To improve the health of people living with SCD, this network conducted activities to increase access to and quality of care, used a shared measurement strategy, and submitted data to the National Coordinating Center (NCC).

2. National Coordinating Center (NCC)

In partnership with HRSA/MCHB, the NCC collaborated with the regional leads to finalize shared measures, align data collection activities, support communication, and receive data from the RCCs. From submitted data, the NCC created reports for HRSA/MCHB. The NCC also was responsible for writing the Report to Congress and a Model Protocol and for gathering resources for a Compendium of Tools and Resources. The NCC was the National Institute for Children's Health Quality (NICHQ).

3. Oversight Steering Committee (OSC)

The OSC was comprised of RCC leads and additional experts who brought specific knowledge, skills, and connections. For the duration of the Program, the OSC met twice a year to give input and assist in making recommendations.

Program Impact

The Program has served well over 25,000 people with SCD – more than a quarter of the SCD population in the U.S. – through 51 clinical sites and 49 community-based organizations. The five RCCs identified over 1,200 Program SCD clinicians in their regions for whom Program funding could lead to improvements in care. These SCD clinicians were all able to prescribe important disease-modifying therapies to SCD patients. Through the Program, providers increased their knowledge to improve care of the SCD population: Over 3,700 attendees completed provider-to-provider telementoring, including specialized COVID-19 seminars.

Three Healthcare Domains

The Report to Congress reflects work conducted to address three healthcare domains. RCCs collected data through an annual provider survey and quarterly medical record review of clinical quality improvement measures.

Healthcare Domain 1: Increase Number of Providers Using NHLBI Guidelines

Highlights and Select Data:

- Showed robust frequency of hydroxyurea (HU) prescription
- Reflected an increased recognition and use of disease-modifying agents, in addition to HU
- Confirmed both the importance of immunizations and that the processes of immunization assessment and delivery must be improved
- Demonstrated higher rates of Transcranial Doppler (TCD) screening than some national findings but recognized that efforts to increase rates should continue
- Identified barriers to care, including systemic bias and racism, and planned activities to address them

Table 1. Areas of Data Measurement and Annual Percentage Over Project Period

DATA MEASUREMENT	AVERAGE PERCENTAGE OVER PROJECT PERIOD[1]
Pediatric Use of hydroxyurea	70.3%
Adult Use of hydroxyurea	57.7%
Pediatric Use of Other Disease-Modifying Therapies	14.1%
Adult Use of Other Disease-Modifying Therapies	24.6%
Pediatric Pneumococcal Immunization	82.0%
Adult Pneumococcal Immunization	63.5%
Pediatric Transcranial Doppler Screening	65.5%

[1] Calculated as the average of each 6-month aggregate percentage from Quarter 1 2019 to Quarter 4 2020.

Healthcare Domain 2: Use of Telementoring and Telemedicine

Highlights and Select Data:

- Demonstrated the ability to leverage the Program infrastructure to expand the knowledge base and comfort level of SCD providers across the U.S.
- Reflected an increase in engaging and training a broad range of providers, both primary care providers and hematology/oncology physicians, nurse practitioners, and physician assistants
- Showed the RCCs' ability to use telementoring to quickly address information and provider education needs emerging from the COVID-19 pandemic

Table 2. Total Count of Provider Attendance in ECHO Sessions Over Project Period

DATA MEASUREMENT	TOTAL OVER PROJECT PERIOD[2]
Total Count of Provider Attendance in ECHO[1] Sessions	3,745

[1] Extension for Community Healthcare Outcomes
[2] Calculated as the total of each 6-month aggregate count from Quarter 1 2019 to Quarter 4 2020. The number is a total count of attendance not the number of unique participants.

Healthcare Domain 3: Implement Strategies to Improve Care

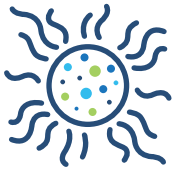
Highlights and Select Data:

- Reflected increased strategies to better meet the needs of patients and families, including coordinating care visits, securing dedicated spaces, co-locating providers, and utilizing multi-disciplinary teams
- Reflected the intentional efforts of RCCs and their CBOs to partner in new and innovative ways to facilitate authentic engagement with patients, families, and caregivers to ensure that they receive comprehensive physical and psychosocial care
- Showed expanded telemedicine offerings which increased access to care for some patients, which were especially important during the COVID-19 pandemic
- Demonstrated improvements in developing and integrating innovative pediatric-to-adult care transition care programs to ensure seamless, comprehensive care at a time when patients are most vulnerable

Table 3. Transition Plan Average Percentage Over Project Period

DATA MEASUREMENT	AVERAGE PERCENTAGE OVER PROJECT PERIOD[1]
Transition Plan for Pediatric-to-Adult Care	40.8%

[1] Calculated as the total of each 6-month aggregate count from Quarter 1 2019 to Quarter 4 2020.



Impact of COVID-19

On January 27, 2020, the U.S. Department of Health and Human Services declared a public health emergency as a result of confirmed cases of 2019 Novel Coronavirus. COVID-19 caused extreme disruption to health care systems throughout the U.S., with many of the providers at local and regional levels in the Program engaged in either front line care or planning and developing procedures and processes to respond to evolving needs. To provide essential care for people living with SCD who were at high risk for complications related to COVID-19, RCCs quickly implemented new workflows and treatment approaches.

In sum, the 2017-2021 Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program addressed clinical and psychosocial needs to improve the health and quality of life of people with sickle cell disease. The Regional Coordinating Centers conducted numerous regional activities in Program priority areas. These activities along with the Program recommendations will inform and enhance future efforts to provide quality care for people living with this complex condition. Details of the Program, including additional data and RCC activities are found in the Report to Congress and its Appendix A, and Recommendations. Additionally, this Report includes a Model Protocol and Compendium of Tools and Resources consisting of resources to improve care of people living with sickle cell disease.

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SICKLE CELL DISEASE TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Report to Congress

SEPTEMBER 2021

Report to Congress

Legislation and Funding Opportunity Announcement

This *2021 Report to Congress* describes the work and data of the current funding cycle of the Sickle Cell Disease (SCD) Treatment Demonstration Regional Collaboratives Program (SCDTDRCP), which was reauthorized by the Sickle Cell Disease and Other Heritable Blood Disorders Research, Surveillance, Prevention, and Treatment Act of 2018, 42 U.S.C. § 300b-5 (2018). This is a continuation of the work the federal government has supported to advance healthcare access and quality of care for those with SCD.

BUILDING ON KEY LESSONS

The work of this Program continued to learn from and build on previous HRSA-funded SCD work. Key learnings have been collected and documented in the Congressional Reports released in 2014 and 2017 and used to inform current work.

The purpose of the Program as outlined in the 2017 Funding Opportunity Announcement was to improve health outcomes in individuals with SCD, reduce morbidity and mortality caused by SCD, reduce the number of individuals with SCD receiving care only in emergency departments (EDs), and improve the quality of coordinated and comprehensive services to people with SCD and their families. The Program was administered by the Health Resources and Services Administration (HRSA) of the Department of Health and Human Services with funding provided through the Maternal and Child Health Bureau (MCHB), Division for Children with Special Health Care Needs. The Program has served well over 25,000 people with SCD – more than a quarter of the SCD population in the U.S. – through 51 clinical sites and 49 community based-organizations. The five RCCs identified over 1,200 Program SCD clinicians in their regions for whom Program funding could lead to improvements in care. These SCD clinicians were all able to prescribe important disease-modifying therapies to SCD patients. Through the Program, providers increased their knowledge to improve care of the SCD population: Over 3,700 attendees completed provider-to-provider telementoring, including specialized COVID-19 seminars.

GLOSSARY TERMS & NOTES ON NAVIGATION

Terms highlighted in red link to the Glossary. To improve the user experience, many cross-document links are in this PDF. In most PDF readers, use the CMD/ALT plus ← (arrow) keys to navigate back to the prior page.

NOTE: Throughout this report, the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program is referred to as "the Program" or "the SCDTDRCP."

Three Healthcare Domains of the Program

As noted in the 2017 Funding Opportunity Announcement, this report reflects data within three healthcare domains:

1. Increase the number of providers treating individuals with SCD using the National Heart, Lung and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report
2. Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care
3. Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

Program funding supported five national Regional Coordinating Centers (RCCs) to establish networks and provide leadership and support for regional and statewide activities to develop and establish systemic mechanisms to improve the prevention and treatment of SCD. HRSA also contracted with a National Coordinating Center (NCC) to assist with data collection. With oversight from HRSA/MCHB, the RCCs, NCC and the Oversight Steering Committee (OSC) collaborated to plan and implement the work of the Program.

Introduction to Sickle Cell Disease

Dr. James B. Herrick first described SCD in the West more than 100 years ago (Frenette & Atweh, 2007). Since then, advances have been made in determining the precise molecular basis for the symptoms and complications of SCD, and establishing screening techniques to identify newborns with the disease (Benson & Therrell Jr, 2010). Although considered a "rare disease," SCD is one of the most common genetic conditions, caused by a single gene mutation that affects the red blood cells (Piel et al., 2017). People who have this mutation can experience a range of symptoms from mild to severe, and those symptoms can change during a lifetime with the disease. The mutation

causes red blood cells to form into the shape of a sickle, with edges of the cells transforming from rounded to sharp. When the "sickled" red blood cells move through blood vessels, they can get stuck.

When the sharp edges press against the walls of the blood vessels, this can cause pain crises. Potent medications have been developed and new therapies are emerging to address complicated symptoms.

Prevalence

Currently, there are approximately 100,000 people with SCD in the United States and millions globally. In the U.S., SCD is most common among African Americans. However other racial and ethnic groups are affected, including Latinos and people of Middle Eastern, Indian, Asian, and Mediterranean backgrounds. Sub-Saharan Africa has the greatest burden of disease.

- Overall, one in nearly 2,000 newborns in the U.S. have SCD (Kato et al., 2018), making SCD the most common condition detected by screening.
- Each year in the U.S., an estimated 1 in 400-500 Black or African-American children are born with SCD (Benson & Therrell Jr, 2010; Feuchtbaum et al., 2012; Kato et al., 2018).
- SCD occurs among about 1 out of every 16,300 Hispanic-American births (Centers for Disease Control and Prevention, 2020b).

Sickle Cell Disease: Severe Physical Complications

In more severe cases, the sickled cells block the flow of blood through vessels, which results in significant illnesses. The most severe physical complications:

Pain Crisis: A devastating hallmark of the disease, pain crisis, also known as **vaso-occlusive pain crisis**, are experienced by nearly all people with SCD, and most will suffer both acute and chronic pain in their lifetime (Payne et al., 2020).

Infection: Damage to the spleen makes people with SCD, especially young children, more susceptible to bacterial infections.

Acute Chest Syndrome (ACS): A condition affecting the lungs that is defined as a new radiodensity on chest radiograph accompanied by fever and/or respiratory symptoms. Damaged lungs lose their primary functionality and are a leading cause of death for people with SCD (Platt et al., 1994).

Stroke: An estimated 10 percent of young children with SCD have a stroke, resulting in diminished cognitive ability (Centers for Disease Control and Prevention, 2020a). Strokes are also a leading cause of death for people with SCD (Platt et al., 1994).

One of the most effective treatments to manage severe and unpredictable pain crises of SCD is opioids. However, long-term opioid use can result in patient health problems and challenges to the healthcare system, which must be acknowledged and addressed. The U.S. has worked to stem the opioid epidemic, but this has made it difficult for some patients living with SCD to get opioid prescriptions filled. Patient experiences of pain are often stigmatized in the healthcare system because of underlying concerns about drug-seeking, though provider underestimation of pain levels has been found to be common (Haywood Jr et al., 2013; National Heart Lung and Blood Institute, 2014; Smith et al., 2008). Coupled with racial bias and structural racism, the barriers to receiving quality, comprehensive, unbiased care remains daunting and results in patients delaying or avoiding seeking ambulatory care for ongoing management of their SCD (Power-Hays & McGann, 2020). Additional information about Program work in this area is in Appendix B. Read more about quality-of-life issues for people with SCD.

The Program Partners: Regional Coordinating Centers, National Coordinating Center, and Oversight Steering Committee

Five Regional Coordinating Centers

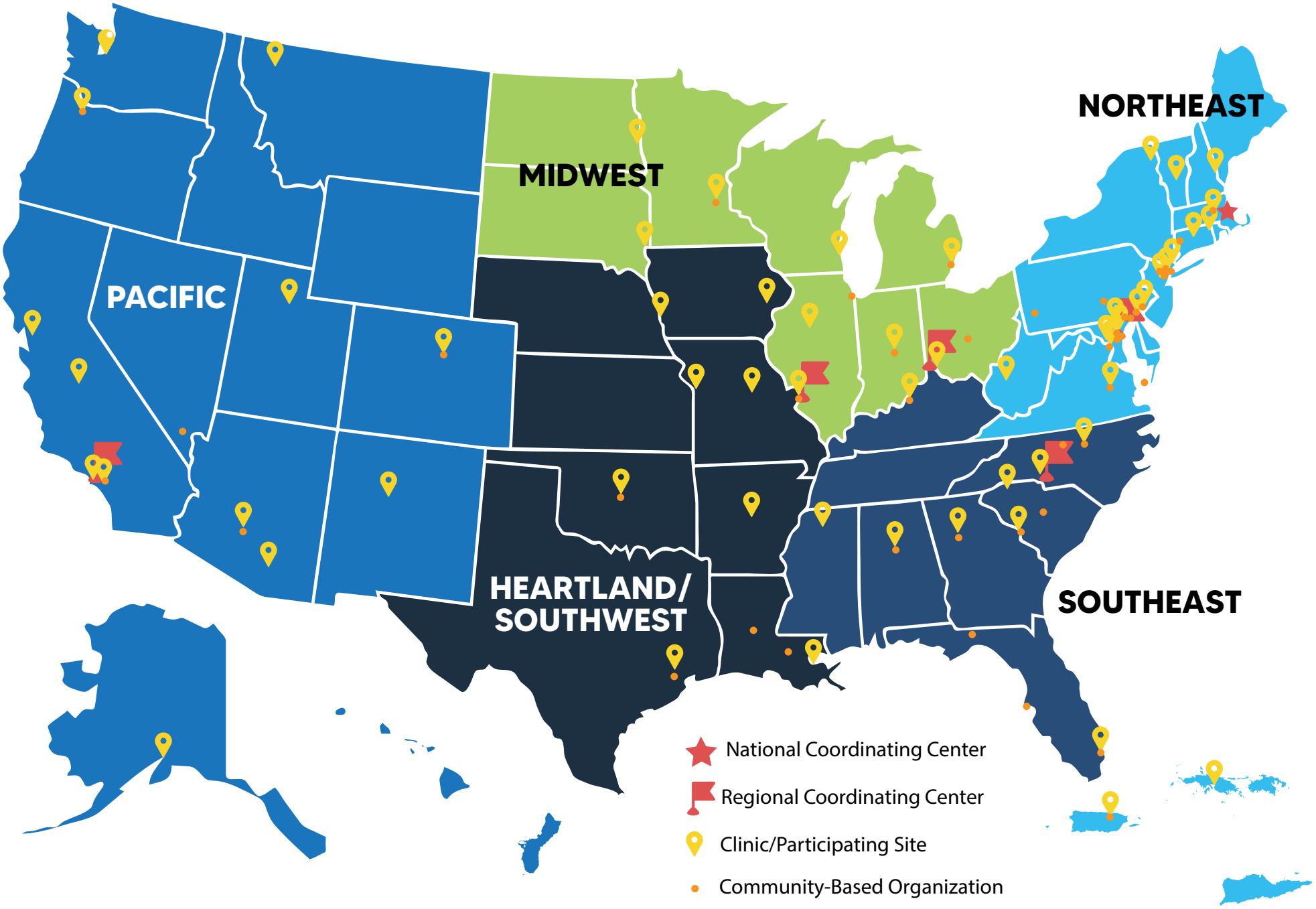
The RCCs established a regional infrastructure and network comprised of clinical locations and community-based organizations (CBOs). Each region had at least one designated regional lead. To improve the health of people living with SCD, this network conducted activities to increase access to and quality of care, used a shared measurement strategy, and submitted data to the National Coordinating Center (NCC). A comprehensive map of Program Partners and details on roles are in Appendix C.

The Program Network

The Program network supported care improvement on a national scale by engaging and supporting smaller, less-resourced sites to tap into the experience of larger participating centers with a greater number of people with SCD and more extensive experience, and by supporting efforts to

SICKLE CELL REGIONAL COLLABORATIVES

Figure 1. Map of National Coordinating Center, Regional Coordinating Centers, Clinics and Participating Sites, and Community-Based Organizations



increase the number of providers who are interested and feel equipped to care for people with SCD. Many providers emphasized how important it was to be part of a broader network of SCD providers for learning opportunities.

The structure and framework of the Program allowed for collaboration, which was particularly useful when specific questions arose. RCC leads could email all regional clinical sites or reach out to the other RCCs to obtain responses for a broader perspective. The ability to get timely, expert input was an important feature of this network. Smaller sites, often in rural, more isolated areas, single sites, or providers serving patients living in large geographic areas can now easily reach providers in the greater RCC networks for problem-solving and case discussions.

National Coordinating Center

In partnership with HRSA/MCHB, the NCC collaborated with the regional leads to finalize shared measures, align data collection activities, support communication, and receive data from the RCCs. From submitted data, the NCC created reports for HRSA/MCHB. The NCC developed this Report to Congress, a Model Protocol, and gathered resources for a Compendium of Tools and Resources. The National Institute for Children's Health Quality (NICHQ) was the NCC.

Oversight Steering Committee

RCC leads and additional experts who brought specific knowledge, skills, and connections comprised the OSC. For the duration of the Program, the OSC met twice a year to give input and assist in making recommendations. The OSC Roster is in Appendix B.

Data Methodology

There were two sources of *quantitative data* collected:

1. Provider Survey for Performance Measurement (PSPM) obtained through an annual online survey. The PSPM reports can be found here.
2. Clinical Quality Improvement Measures (CQIMs) obtained through quarterly review of medical records (electronic or manual). The Q1 2019 through Q4 2020 CQIM reports can be found here.

Qualitative data were obtained by key informant interviews and gathering of reports and presentations shared in meetings among the RCCs. See Table 1 for the measures collected based on the Program objectives. Appendix C has full detail of the data methods, inclusion/exclusion criteria, and annotations.

Table 1. Provider Survey for Performance and Clinical Quality Improvement Measures

PROVIDER SURVEY FOR PERFORMANCE MEASURES (PSPM)				
COLLECTED ANNUALLY (2019, 2020)				
ELECTRONIC OR PAPER SURVEY SENT TO PROVIDERS				
1. Number of providers in the SCDTDRCP	**2.** Number of patients seen by a SCDTDRCP provider in the past year	**3.** Number of providers in the SCDTDRCP participating in telementoring for SCD in the past year	**4.** Number of SCDTDRCP providers who report feeling comfortable treating people living with SCD	**5.** Number of SCDTDRCP providers who saw at least one patient in the past year and who prescribed hydroxyurea (HU)
CLINICAL QUALITY IMPROVEMENT MEASURES (CQIM)				
COLLECTED QUARTERLY** (2019-2021)				
EMR DATA PULL OR MANUAL CHART REVIEW COMPLETED				
1. Hydroxyurea (HU) Use: measured by prescription rates (REQUIRED)	**2.** Transcranial Doppler (TCD) screening	**3.** Immunizations	**4.** Transitions in Care	**5.** Project ECHO® (provider-to-provider telementoring)

Note: All RCCs were required to collect HU use data. They also were required to select at least one additional measure.
* Four of the five RCCs collected data quarterly. The fifth RCC collected data every six months.
+All sites that initiated data collection may not have submitted data every quarter of the Program.

The data presented for the Program were collected for Quality Improvement (QI) purposes. In QI work, data are used to guide the implementation of activities; accordingly, data should be considered in the context collected.

Interpretation of the data should consider the following:

- The data were collected through convenience sampling.
- Measure denominators fluctuated over time due to variation in sites reporting each quarter.
- The COVID-19 pandemic coincided with the 2020 data collection period.

Given these factors, definitive assessment of improvements in program objectives across time is not possible and findings are not fully generalizable.

IMPACT OF THE COVID-19 PANDEMIC

On January 27, 2020, the U.S. Department of Health and Human Services declared a public health emergency because of confirmed cases of 2019 Novel Coronavirus. COVID-19 caused significant disruption to health care systems throughout the U.S., and many of the SCD providers in the SCDTDRCP were engaged in either front line care or planning and developing procedures and processes, or both, to respond to evolving needs. Healthcare systems and individual providers quickly implemented new workflows and approaches to provide essential care for people living with SCD – who were at high risk for serious complications of COVID-19. Appendix B contains additional information about the impact of COVID-19 on people living with SCD and on the Program.

Healthcare Domain 1

Increase the number of providers treating individuals with SCD using the National Heart, Lung and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report

Domain 1 Highlights

- Robust frequency of HU prescription: On average, 70% of the pediatric population and 58% of the adult population served by this program received an HU prescription*
- Increasing recognition and use of disease-modifying agents, other than HU: On average, 14% of the pediatric population and 24.6% of the adult population served by this program received disease-modifying therapies (other than HU)*
- Confirmation of the importance of immunizations and acknowledgment that the processes of immunization assessment and delivery must be improved: On average, the pediatric pneumococcal immunization rate was 82% and the adult pneumococcal immunization rate was 63.5%*
- Demonstration of higher rates of TCD screening than some national findings, but recognition that efforts to increase rates should continue. The average pediatric TCD rate was 65.5%*
- Identification of barriers to care, including systemic bias and racism, and planning activities to address them

*Calculated as the average of each 6-month aggregate percentage from Quarter 1 2019 to Quarter 4 2020.

RCCs collected information in three clinical areas:

Hydroxyurea (HU) prescribing, a disease-modifying medication effective in reducing pain crises and decreasing hospitalization and ER visits

Immunization, a prophylactic therapy that prevents life-threatening infections, with specific focus on pneumococcal vaccination

Transcranial Doppler (TCD) screening, a procedure that identifies children who are at higher risk for stroke

Importance of Hydroxyurea Use and Other Disease-Modifying Therapies for Individuals with SCD

Hydroxyurea has been shown to significantly reduce the frequency of SCD-related pain, need for blood transfusions, and common pulmonary complications, including ACS (National Heart Lung and Blood Institute, 2014). Left untreated, ACS has been shown to cause significant morbidity and is associated with a higher risk of death. HU has been approved by the U.S. Food and Drug Administration (FDA) for the treatment of clinically severe SCD since 1998 for adults and since 2017 for children. Additional HU information and NHLBI guidelines for HU use can be found in the Recommendations. However, despite the strong NHLBI recommendation, uptake has been inconsistent and below recommended levels (Brousseau et al., 2019; Su et al., 2019). A discussion of factors that may contribute to low rates of HU is in Appendix B.

The Program assessed adherence to HU use recommendations by 1) reviewing health record information to measure the number of patients given a prescription for HU (CQIM, see Table 2 and Table 3); and 2) surveying providers to determine the percentage who report prescribing HU (PSPM). Results demonstrated high, consistent support of the use of HU among participating sites and higher percentages of HU usage compared to other available data on the national experience. Every RCC provided CQIM data on HU utilization within their regions. On average, 70 percent of the pediatric population served by this program received an HU prescription and 58 percent of the adult population served by this program received an HU prescription.

Table 2. Hydroxyurea Use Aggregated into Six-Month Increments for Pediatric Population Treated at RCCs and Participating Sites

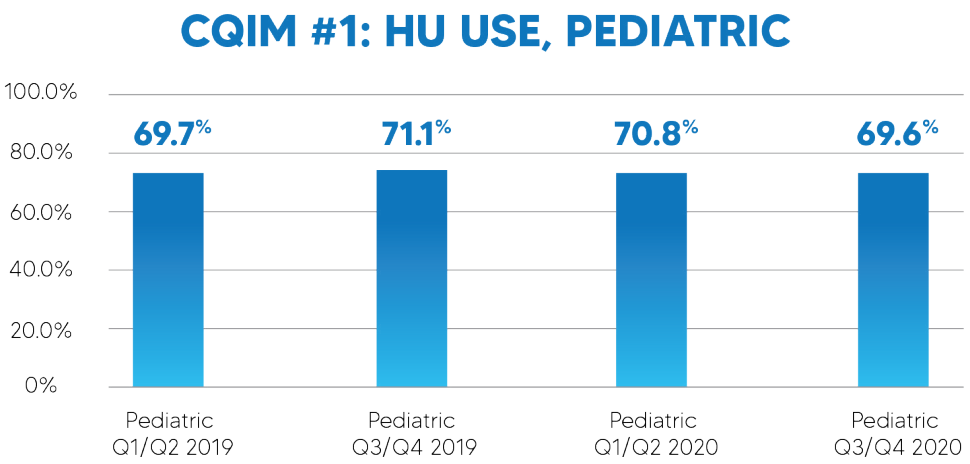
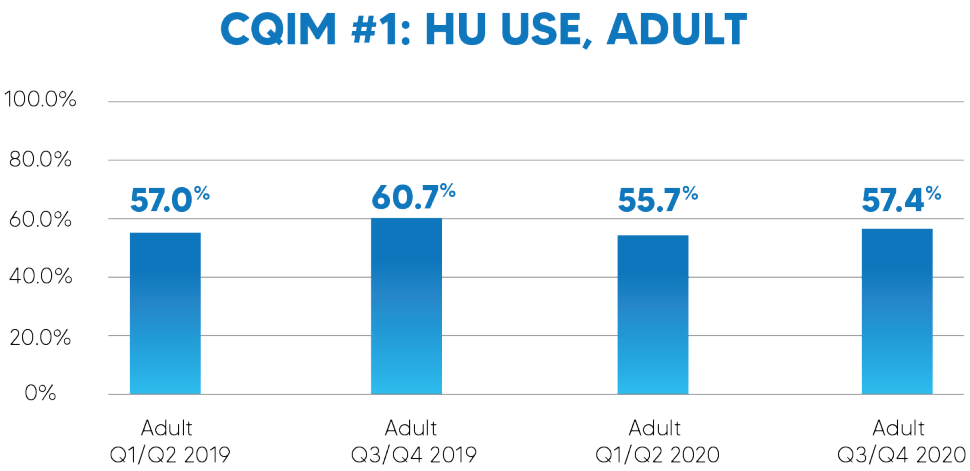


Table 3. Hydroxyurea Use Aggregated into Six-Month Increments for Adult Population Treated at RCCs and Participating Sites



Data from the Program's PSPM also suggest high utilization of HU:

- In 2019 and 2020, between 18,000-20,000 patients out of approximately 26,000 total patients seen (both adult and pediatric populations) were given an HU prescription in the last year, with an average of 75 percent of the pediatric patients and 67 percent of adults given a prescription.
- Almost 50-65 percent of providers surveyed reported prescribing HU in the past year (YR1 47.7 percent; YR2 65 percent).

RCC HU-Focused Work

Further assessment is needed to fully explain the relatively high utilization of HU use compared to national findings. Program work completed to develop infrastructure to track HU use and reduce burden and barriers associated with ordering a prescription may be a contributing factor. Several of the Program sites specifically programmed their Electronic Health Record (EHR) systems to track clinical care elements, including HU prescription, so that accurate counts of treatments can be obtained quickly. In addition, some sites instituted QI projects to improve the tracking of HU and, in turn, increase its use. Because this type of analysis is time- and resource-intensive, QI activities are not currently feasible for all programs. Additional discussion of national data, barriers to HU use and prescription, factors that may have contributed to the Program HU use, and examples of RCC activities can be found in the Appendix.

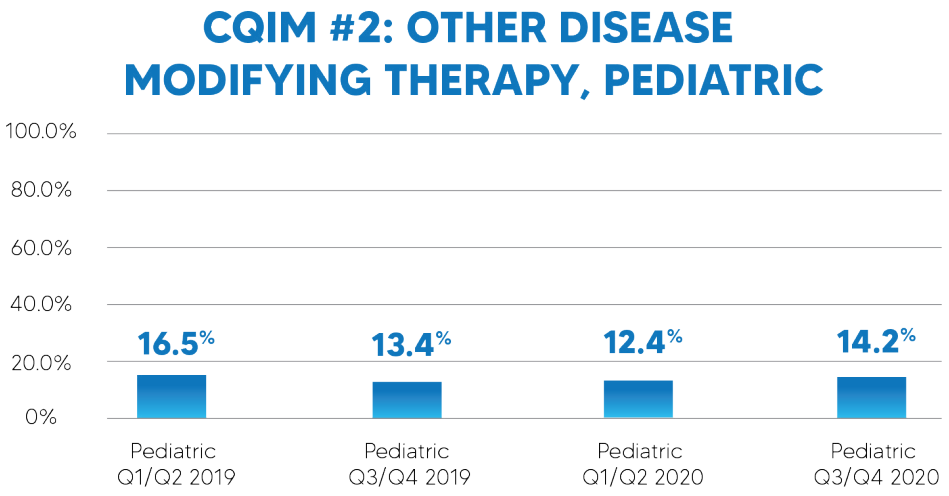
Two RCCs described QI projects to track when discussions are completed with patients about therapies and decisions to modify treatment plans. The tracking documents patient responses during discussions; if the patient or caregiver is turning down treatments, the site flags and reviews the notes to better understand the decision-making process and concerns so that the clinical team can provide the right information to the patient. Sites have found that patients and caregivers may need more information about the treatment and its side effects to dispel myths and clarify explanations. These include print or electronic resources; referrals to connect with families who decided to place their child on disease-modifying therapies; lived experiences of older patients on specific treatments; support from CBOs; and continued discussion at each clinic visit.

Beyond HU: Other Disease-Modifying Therapies

Most people with SCD meet the criteria for HU prescription. Providers in the Program agree that HU should continue to be one of the standards of care. However, because some patients have reached their maximum dosage or cannot take HU for other reasons, additional therapies are needed. RCCs are actively participating in research in this area, such as trials related to bone marrow transplantation and other alternative disease-modifying therapies. Transplant is currently the only cure for SCD and sites are trying to expand this treatment, though it is costly and requires donor matching that makes it uncommon. Additional information about bone marrow transplant is in Appendix B. Emerging and novel therapies will have a broader role in treatment options offered either in combination with HU or alone,

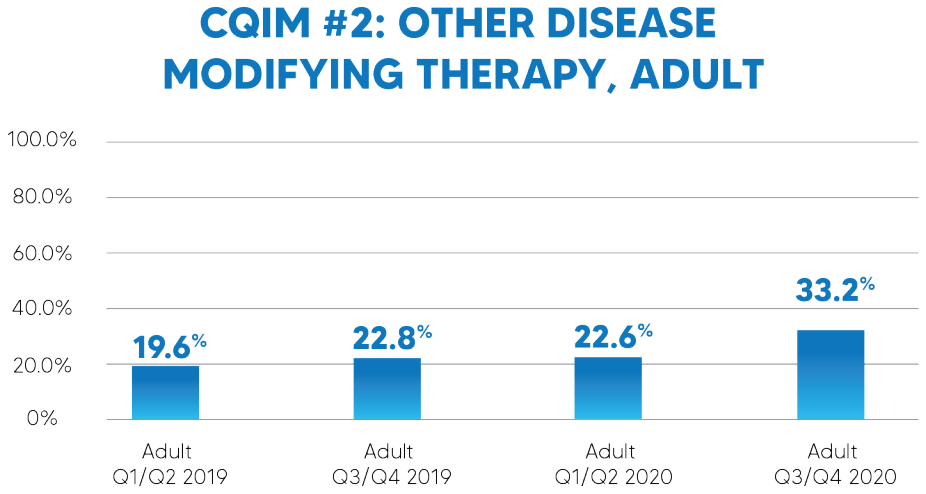
including: **Adakveo (crizanlizumab)**, **Oxbryta (voxelotor)**, and **Endari (L-glutamine)**; and the use of **erythrocytapheresis**, a non-surgical treatment red cell exchange transfusion. Additional information about these emerging medications and advances in gene therapy can be found in the Appendix. With increased options, using combination therapies may be the solution to address the myriad health issues faced by people with this complex disease. RCCs have increased tracking the use of disease-modifying therapies other than HU. During the time of the Program, they elected to collect initial data about these other therapies though they were not a focus of the original measurement strategy. Data collected from the Program suggests some growth in utilization of disease-modifying therapies beyond HU among the adult population but use in pediatric populations is still emerging. The results from these data are modest and should be viewed only as an initial assessment. Continued work is needed to capture more information about specific types of therapies used and frequency of use at treatment sites.

Table 4. Disease-Modifying Therapy Use Other Than HU Among Children/Adolescents



Percent of patients ≥9 months and <18 years of age prescribed disease-modifying therapy other than HU.

Table 5. Disease-Modifying Therapy Use Other Than HU Among Adults



Percent of patients 18 years and older prescribed disease-modifying therapy other than HU.

Immunizations

People with SCD are at increased risk for invasive bacterial diseases, like **pneumococcal disease**. The NHLBI guidelines recommend that all patients, unless otherwise advised, be immunized based on the CDC's Advisory Committee on Immunization Practices. Still, comprehensive rates are subpar. Appendix B contains immunization guidelines and national rates.

Immunization Data Collection Challenges

Although all RCCs strongly supported the routine and consistent administration of preventive immunizations, access to comprehensive immunization records were not available to many program sites, particularly subspecialty clinics without access to statewide immunization registries. This made data collection challenging. Additional discussion of the Program immunization data collection challenges is in Appendix C. Thus, during the Program, several teams focused on both the data collection process (access to data and data quality) and actual immunization utilization.

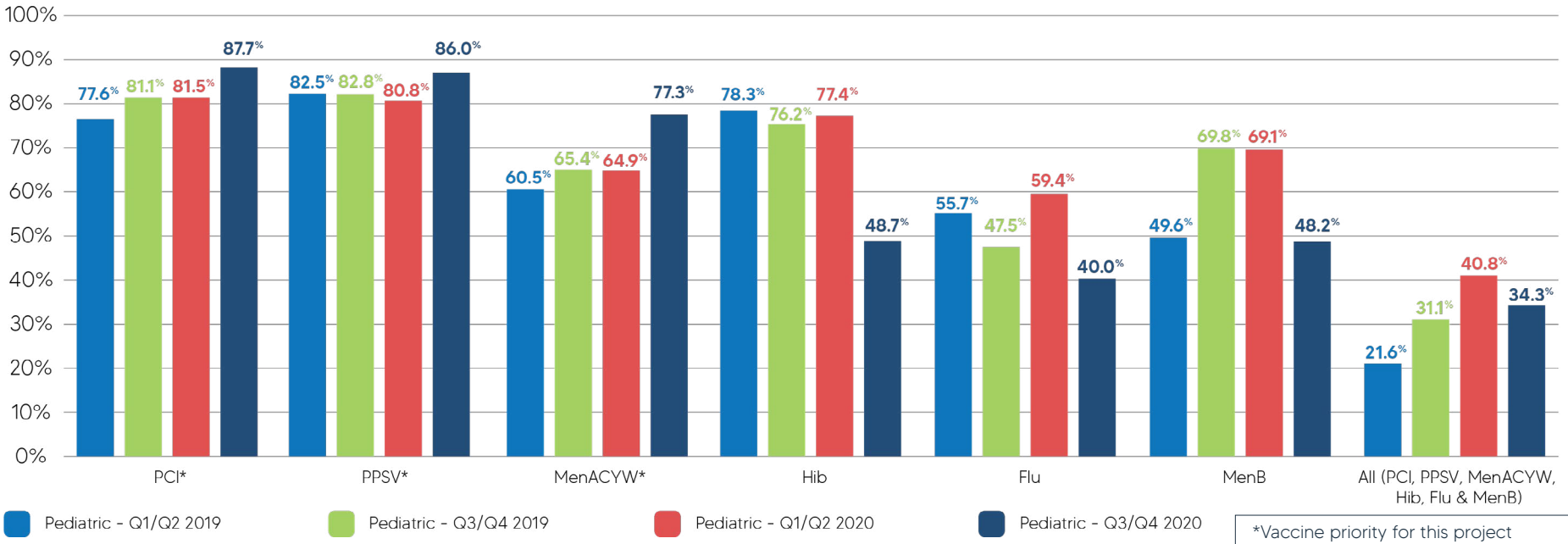
Tables 6 and 7 show information from the sites that reported immunization data. It is important to note that reporting across quarters was inconsistent, so these data should not be considered population-level information but rather select clinic representation. Although the overall results may be higher due to missing data, trends toward improving immunization coverage during the Program can be seen for many of the vaccine series.

RCC Immunization-Focused Work

To improve immunization data collection, several RCCs created elaborate dashboards, managed clinical **EHR systems**, or manually maintained spreadsheets to track immunization data. Pneumococcal Immunization (PCI) was a specific priority area for the Program, with some participating sites focusing on PCI in QI efforts. The PCI-specific data from the Program suggests increasing PCI rates across both pediatric and adult populations.

Table 6. Immunizations Aggregated by Six-Month Increments for Pediatric Population Among RCCs and Participating Sites

SICKLE CELL DISEASE QI MEASURE 4 | PEDIATRIC IMMUNIZATIONS



*Vaccine priority for this project
PCI = pneumococcal immunization
PPSV = pneumococcal polysaccharide vaccine
MenACYW = vaccine for 4 strains of the meningococcal bacteria – A, C, W, and Y
Hib = Haemophilus influenzae type b vaccine
Flu = influenza (quadrivalent) vaccine
Men B = Meningococcal B vaccine

Table 7. Immunizations Aggregated by Six-Month Increments for Adult Population Among RCCs and Participating Sites

SICKLE CELL DISEASE QI MEASURE 4 | ADULT IMMUNIZATIONS

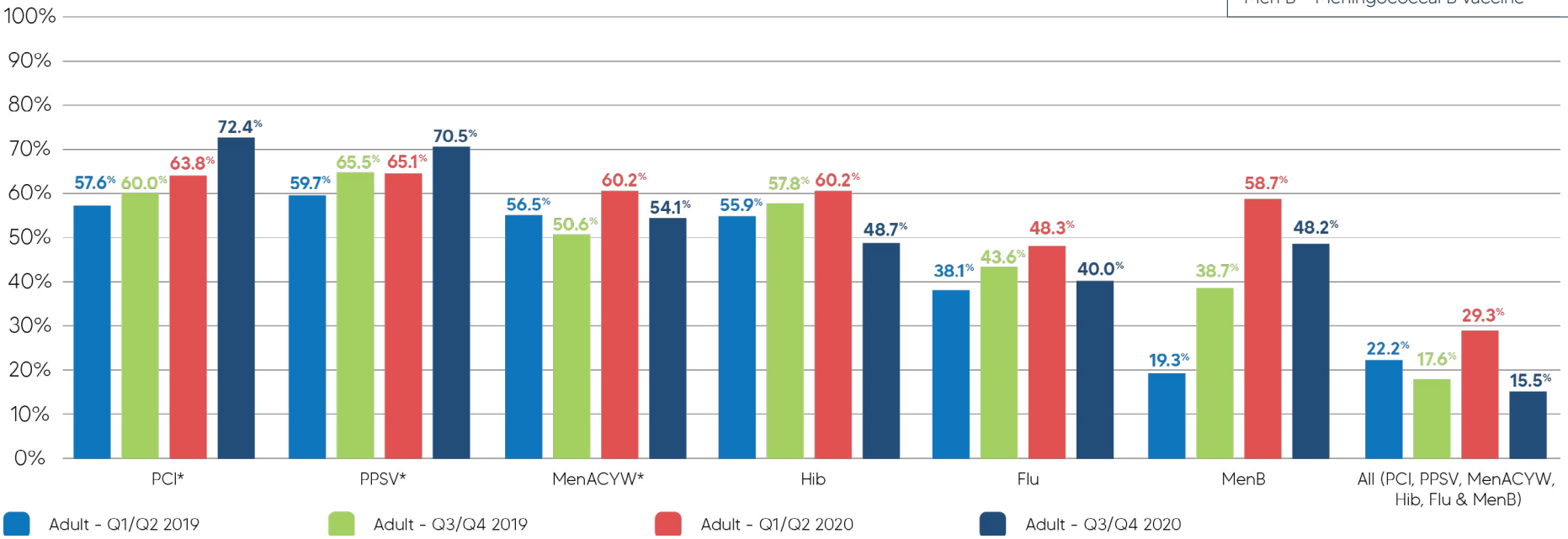
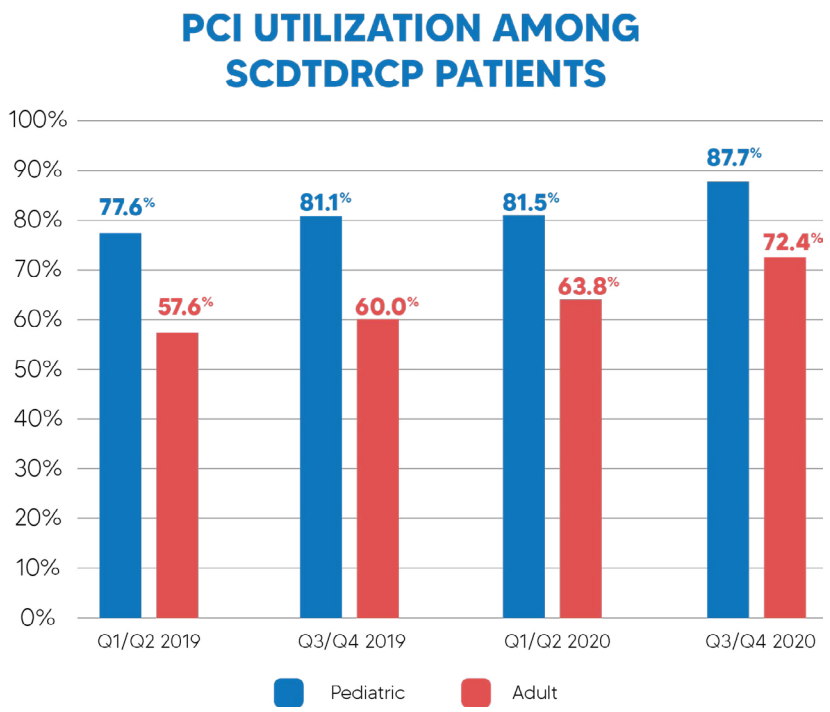


Table 8. CQIM Pneumococcal Immunization Aggregated by Six-Month Increments for Adult and Pediatric Population Among RCCs and Participating Sites



Transcranial Doppler Screening

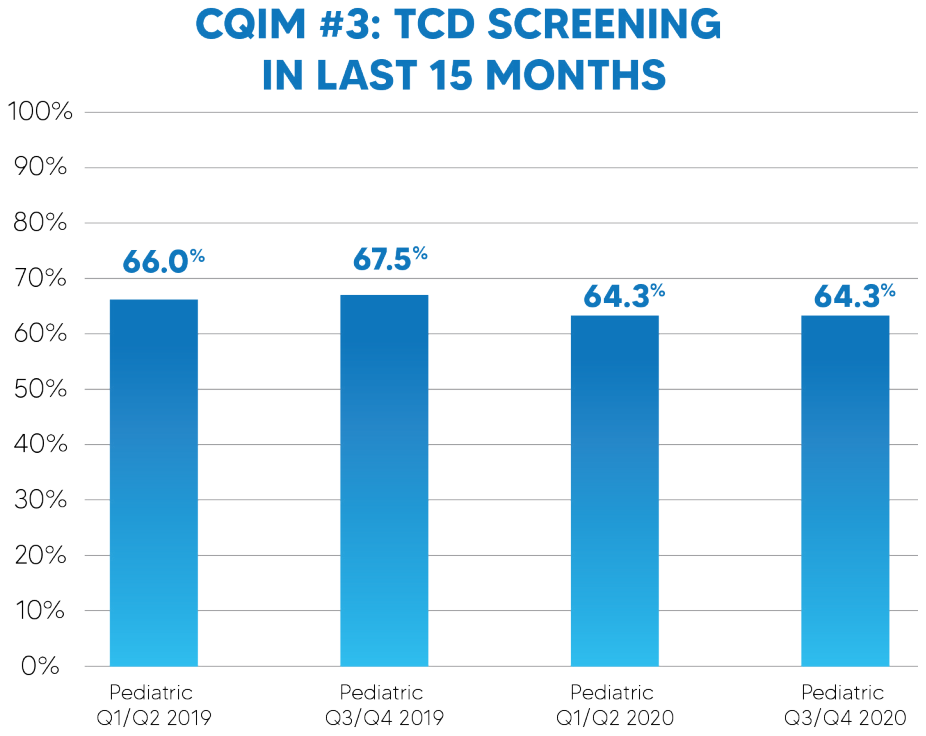
People who live with SCD are at increased risk for stroke, both overt and silent. Adults who experience stroke have severe morbidity and high mortality rates. Transcranial Doppler (TCD) is a noninvasive ultrasound procedure that allows the clinician to clearly see how quickly blood is flowing through the brain over a set time. High blood flow is associated with an increased risk of stroke. The test is reliable, painless, and relatively inexpensive. While HU has helped reduce strokes, programs continue other methods to reduce stroke risk. Given these factors, per the NHLBI guidelines, TCD use with children aged 2-16 years is strongly recommended. Even so, it appears that less than half of eligible pediatric patients receive appropriate TCD screening (National Academies of Sciences Engineering and Medicine, 2020).

RCC Transcranial Doppler-Focused Work

The RCCs conducted extensive work to increase the use of TCD. During the Program, several sites implemented changes to their clinical flows and other processes to ensure that all eligible patients are scheduled for this life-saving preventive screening. Some RCCs implemented QI projects that tracked a patient's last TCD to flag when they would be due again. This type of tracking has been essential in ensuring that programs do not allow more than a year between TCDs. One region is leading a comprehensive effort to create a "stroke-free generation." Examples of other initiatives undertaken by RCC sites to improve TCD rates are in the Appendix. However, there are some serious barriers to care that RCCs continue to address in this area.

The myriad ways that RCCs and sites have focused work on the clinically supported guidelines for TCD appear to have led to progress, as data collected for the Program shows moderately high percentages of TCD completion. On average, 65 percent of eligible patients aged 2-16 years had a TCD screening in the last 15 months, as shown in Table 9.

Table 9. CQIM Transcranial Doppler Screening Completed Within the Last 15 Months for Patients Aged 2-16 Years in Six-Month Increments Among RCCs and Partner Sites



Additional Areas of RCC Focus on Evidence-Based Care

Much of the work described in this report focuses on the clinical areas of HU, immunizations, and TCD. However, the RCCs conducted important work in several other areas covered by NHLBI report, including – and especially – management of pain crises.

Pain Management

Pain crises are a hallmark of SCD and the NHLBI 2014 Evidence-Based Management of Sickle Cell Disease report has several robust clinical recommendations regarding pain management (National Heart Lung and Blood Institute, 2014). People with SCD who present in pain to emergency departments (EDs) and other clinical settings need immediate attention. Because of the particular importance of addressing and improving this area of care, the Program has developed a recommendation about ED care and this report provides a deeper look into activities of the RCCs and the challenges of managing pain crises, especially given the opioid epidemic.

Impact of Bias on People with SCD

All RCCs discussed the impact that systemic bias has on their patients. At the patient level, there is inequitable access to care, unequal attention to treatment development compared to other types of conditions, and significant bias – especially as it relates to obtaining timely and appropriate pain care. As noted by the National Academies of Science, Engineering, and Medicine SCD (National Academies of Sciences Engineering and Medicine, 2020), when discussing SCD care and the people with SCD, it is essential to point out that stigma, implicit bias, and racism are key societal factors that contribute to the burden of disease. Pain management of SCD is impacted by these factors, especially those that exist in patient and provider interaction. Addressing and decreasing the impact of these factors is essential to increase provider comfort and overall improvement in SCD care when treating this population. RCC activities to address this issue included: conducting hospital-wide implicit bias training; discussing cases that demonstrated unwarranted differences in care; and addressing the issue directly by educating providers and confronting bias. Appendix A contains examples of RCC activities related to addressing bias.

Healthcare Domain 2

Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care

Domain 2 Highlights

- Leveraging the Program infrastructure and support built through state plans, teaching opportunities, and provider-to-provider communication, RCCs expanded the knowledge base and comfort level of providers across the U.S. with this program, improving access to care for people living with SCD.
- Engaging and training a broad range of providers, both specialist and non-specialist (e.g., primary care and hematology/oncology physicians, nurse practitioners, and physician assistants) using Project ECHO® and other education opportunities to increase knowledge among providers. Topics ranged from disease-modifying therapies, psychosocial issues, insurance barriers and practice guidelines. This resulted in more than 3,700 attendees of telementoring, which strengthened care locally and nationally.
- Using telementoring quickly addressed emerging information and provider education needs caused by the challenges of the COVID-19 pandemic.

Program Providers

Through the PSPM, RCCs assessed care delivered by providers who care for this population. The data obtained provides insight into the composition of providers who worked closely with the RCCs and responded to the survey. *When reviewing these data, consider the following: the pool of providers who were sent and answered the survey each year varied and the COVID-19 pandemic was occurring during the second survey.*

Overall, the data reflect and highlight national trends:

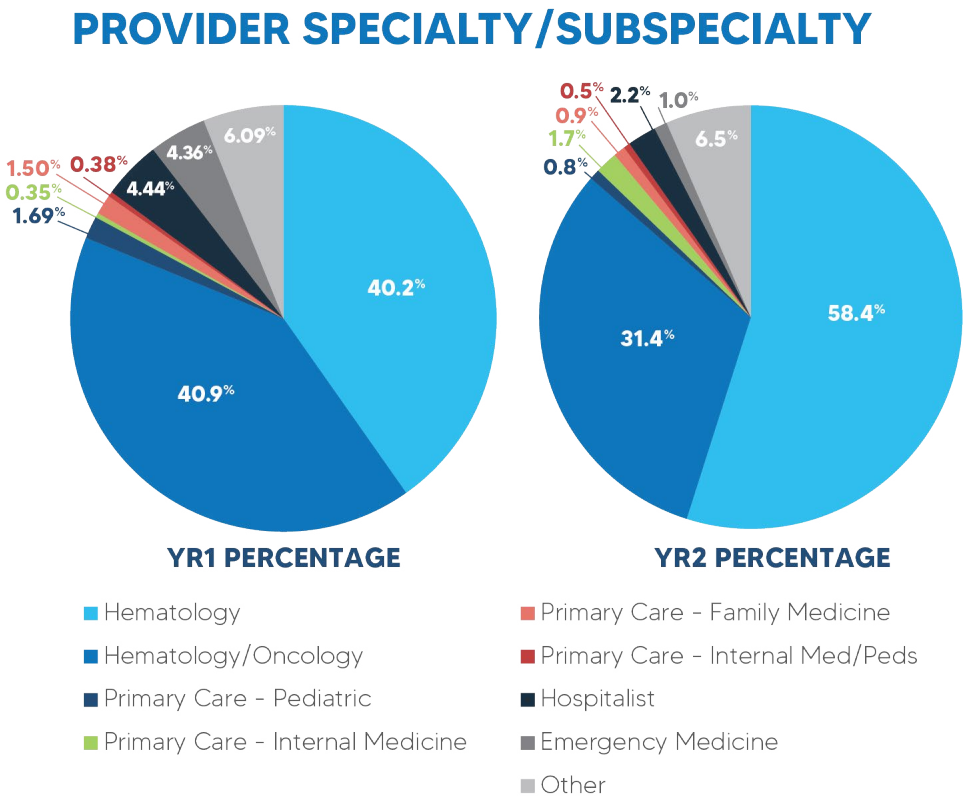
- A relatively small subset of clinicians provide the vast majority of care for people with SCD
- Most SCD care is provided by specialists (hematologists) rather than PCPs and other generalists
- Few SCD care providers are located in rural settings

RCCs initiated the following surveys: In YR1, 1,854 providers were sent the PSPM. Using a refined definition of SCD provider inclusion, in YR2, 1,220 providers were sent the PSPM. The total number of responding providers YR1: 516. The total number of responding providers YR2 was: 306.

PSPM findings include:

- Majority of providers treating patients are medical doctors within the Program: YR1 406/516 (78.7%); YR2 236/306 (77.1%)
- Approximately one-third (1/3) of providers in the Program only see adult patients, one-third (1/3) only see pediatric patients, and the remaining one-third (1/3) see both
- Specialists comprised the majority of providers:
    - Y1: Hematologists: 10,885/27,078 (40.2%); Hematologists/Oncologists: 11,082/27,078 (40.9%), leading to a total percentage of 81.1 percent
    - Y2: Hematologists: 14,081/25,712 (54.8%); Hematologists/Oncologists: 8,071/25,712 (31.4%), leading to a total percentage of 86.2 percent
- Nearly all clinicians affiliated with the RCCs were in an urban setting: YR1 497/516 (96.3%); YR2 277/306 (90.5%) and 50 percent or more worked in a practice affiliated with a university or medical school YR1 256/516 (49.6%); YR2 194/306 (63.4%)

Table 10. Percentage of Patients Seen by Each Practice Specialty (PSPM#2)



Nationally, there is a pressing need for more providers who are willing and adequately prepared to care for people with SCD, especially adult care providers. Lack of adult providers is particularly concerning to pediatric specialists who must rely on this next group of providers during the important time of transition to adult care. Deeper discussion of need for providers is in Appendix B. While the limited number of hematologists is a focal point, other providers, including radiologists, pulmonologists, cardiologists, orthopedics, nephrologists, social workers, psychologists, and supportive care providers who understand SCD are also needed and in limited supply. RCCs noted these shortages and how it negatively impacts adequate, timely care.

The Program included two areas of work to increase provider knowledge and to support training providers to follow NHLBI care guidelines: 1) knowledge-sharing, and 2) improving comfort and confidence level of providers who may care for people with SCD.

Knowledge-Sharing

Knowledge-sharing between trusted colleagues is an effective way of increasing awareness. The Program offered education and training support to increase the education of providers, clinicians, and staff improving the lives of patients. This section describes the ways RCCs, states, and participating sites sought to share information and expertise during the Program.

Sharing State Plans

As part of the Program, RCCs developed a Regional Sickle Cell Action Plan and state-specific Sickle Cell Action Plans for funded states. The plans included: 1) resources in each region and state to improve SCD care for all people with SCD in the region, and 2) a description of the overall infrastructure that would address the goals and requirements listed in the Funding Opportunity Announcement.

The state action plan described:

- How each state intended to develop a network of providers using evidence-based SCD care in the state
- How telemedicine and telehealth strategies and other provider support would be utilized
- How access to quality care would be supported
- How the state intended to increase the number of individuals with SCD being treated by providers using evidence-based SCD care

These plans were used to help formulate a cohesive approach to facilitate knowledge-sharing. Discussions spurred by these state plans brought forth a regional vision and strong partnerships with the RCCs that continues today. Information included in state plans may be helpful to states interested in doing similar work. Appendix B contains two full plan examples.

Provider-to-Provider Communication

RCCs conducted provider-to-provider education to increase provider capacity. Given the need to build the next generation of pediatric and adult SCD providers, the Program providers spent time in discussions, fielding questions, and sharing their individual expertise with other providers. They also dedicated time formally, including conducting grand rounds, giving educational seminars, and using Project ECHO® (Extension for Community Healthcare Outcomes) to educate residents and physicians across disciplines.

Telementoring with Project ECHO®

During the past decade, telementoring has proven to be a highly successful approach to supporting clinicians remotely, and Project ECHO®, created by Sanjeev Arora, MD, at the University of New Mexico, was an important tool for RCCs to expand training and professional education in the Program. More information about Project ECHO® and how it works is in Appendix B. The Program has developed a recommendation about telementoring.

RCC ECHOs

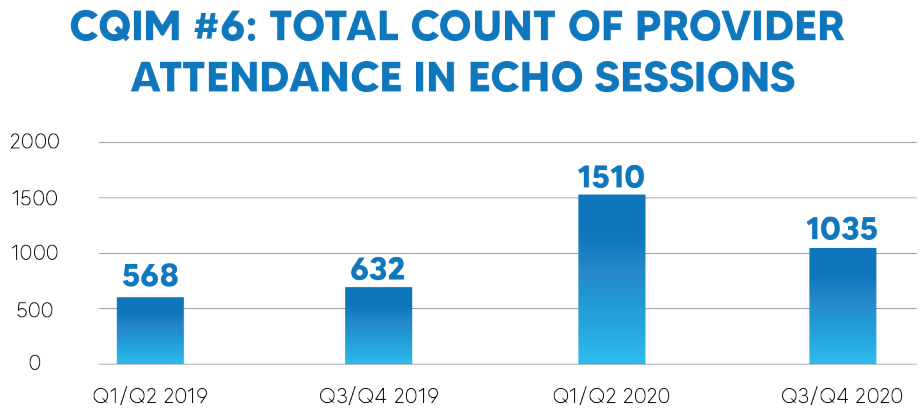
RCCs found the Project ECHO® model effective and manageable for knowledge-sharing, reporting that it helped form a larger community for participants to engage and exchange information. Expert providers within the Program network shared their expertise with attendees. RCCs supported a general regional ECHO, while some also developed topic specific ECHOs, including ones to address emerging COVID-19 needs. A description of RCC activities using the Project ECHO® model is in Appendix A.

The Program collected data on ECHO participation in the CQIM and PSPM.

CQIM

RCCs reported quarterly counts of total number of providers attending telementoring ECHO sessions via the CQIM. Over the four time periods of data collection reflected, the total number of attendees of ECHO sessions was 3,745. Counts include all sessions a provider attended; attendees could have been counted more than once if they attended multiple sessions in the data collection time frame. Counts were aggregated into 6-month increments. See Table 11.

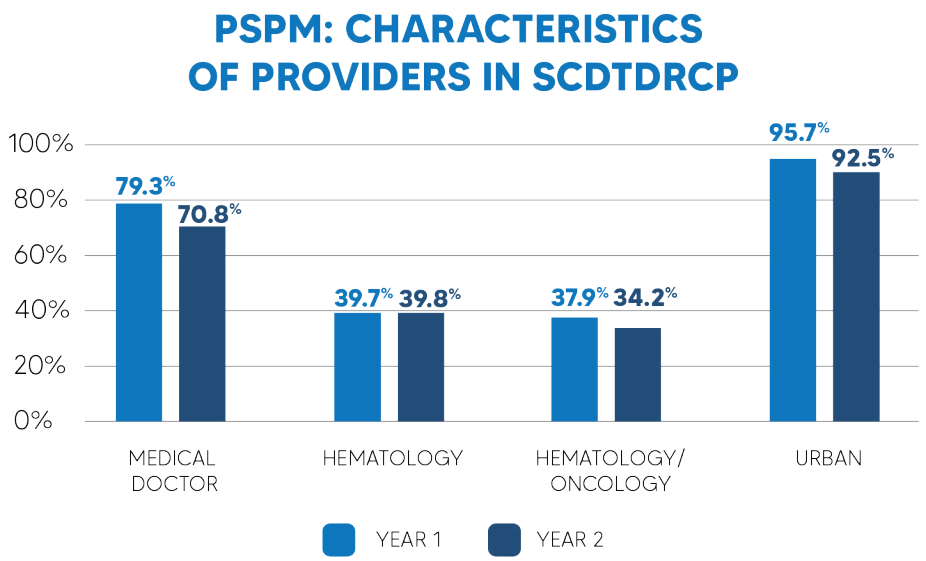
Table 11. CQIM: Providers Participating in the Program ECHO Telementoring Sessions in Six-Month Increments



PSPM

PSPM data shows that 116 individual Program providers who completed the survey question reported participating in telementoring on the first survey and 161 on the second survey (data not shown). About one-third of participating providers reported that they were regional or state leads. Across both survey years, the majority of providers who attended telementoring sessions were medical doctors, hematologists, or hematologist/oncologists, serving in urban areas. See Table 12.

Table 12. PSPM: Characteristics (Provider Type, Specialty, and Location) of SCDTDRCP Providers Who Individually Participated in Telementoring Sessions in the Past Year by Survey Year Among RCCs and Partner Sites



Addressing Provider Comfort Treating People with SCD

National data show that many general providers are not comfortable treating people with SCD, demonstrating the need for multiple outlets for knowledge-sharing. According to a national survey of family physicians, only 20 percent of providers were comfortable treating people with SCD (Mainous et al., 2015). Those who had at least one patient with SCD were significantly more comfortable managing patients, and more than two-thirds of those surveyed responded that they would be willing to co-manage a patient with SCD alongside a specialist.

Like the education opportunities with specialists described in this report, a structure that encourages discussion of unfamiliar clinical scenarios and treatment options with experts who can give timely and direct advice may allow colleagues to feel more comfortable treating patients with SCD.

The data collected for the Program showed that comfort level of providers varied, with specialists showing the greatest comfort. The majority of providers in the Program report feeling comfortable treating SCD patients (YR1- 57.6%; YR2 – 72.2%). Importantly, however, high levels of provider comfort treating patients with SCD is not seen in other research. Being affiliated with a group of knowledgeable, supportive colleagues may be an important protective factor.

The Recommendations section of this report calls for ongoing provider education and practical experience as important aspects of building provider comfort. Continuing to offer experiences via the Program and developing and providing other clinical opportunities should be considered. The Program also recommends co-management of patients when feasible or necessary.

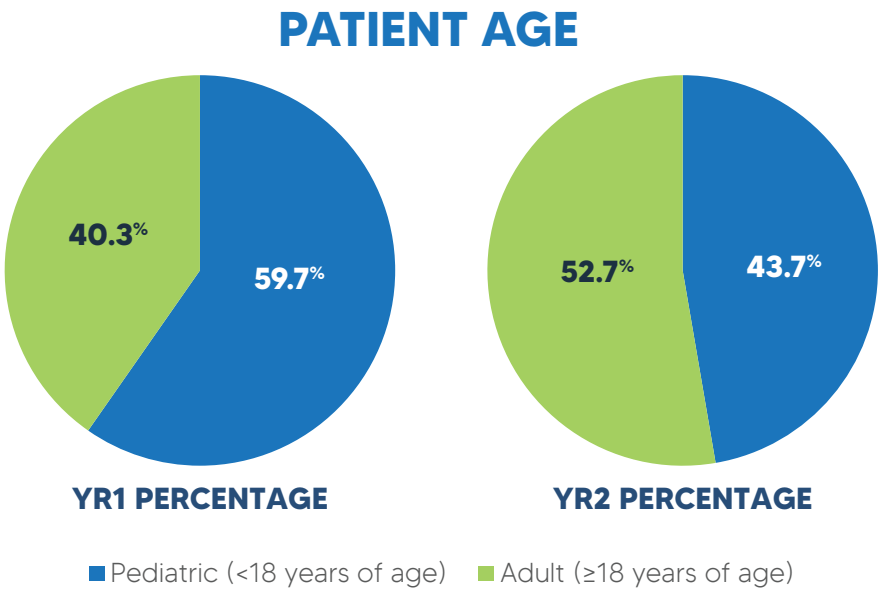
Healthcare Domain 3

Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement and partnership, adolescent transitions to adult life, and care in a medical home

Domain 3 Highlights

- Data collected through the PSPM showed that over 25,000 patients were seen by a Program provider in the past year. Patient ages, of the providers who answered the survey, are represented in Table 13.
- Strategies were implemented to better meet the needs of patients and families, including coordinating care visits, securing dedicated spaces, co-locating providers, and utilizing multi-disciplinary teams.
- RCCs and their CBOs partnered in new and innovative ways to ensure authentic engagement with patients, families, and caregivers to assure comprehensive physical and psychosocial care.
- Expanding telemedicine increased access to care for some patients, which was especially important through the COVID-19 pandemic.
- RCCs and their networks devoted time to developing and integrating innovative pediatric-to-adult transition care programs to ensure seamless, comprehensive care at a time when patients are most vulnerable.
- Specific program activities for this domain are reflected in Appendix A.

Table 13. Age of Patients Seen by a Program Provider (PSPM#2)



Comprehensive, Coordinated, and Continuous Care

Quality care is comprehensive, coordinated, and continues through the lifespan. A medical home is when a practice-based care team takes collective responsibility for a patient's ongoing care (Medical Home Initiatives for Children With Special Needs Project Advisory Committee, 2002). The care team may be co-located, but this is not essential. The most important aspect is that the team is responsible for providing and arranging all the patient's health care needs. The Program has developed a recommendation for coordinated care. Many RCCs expressed interest in moving toward a medical home model and some sites had success but accomplishing medical homes for all people living with SCD is a work in progress at many sites. Refining the process, building relationships, establishing communication streams, and making responsive data systems takes time and resources.

Comprehensive Care

As RCCs worked toward the medical home model, sites approached ensuring that patients were receiving comprehensive care in a variety of ways. For additional information on this topic, see Appendix B.

Sites conducted activities to provide patients with access to comprehensive care. For example, one site implemented the American Society of Hematologists *Patient Summary Review* annually, which ensures that patients are directly asked about their care and prompted about important clinical milestones. Another site moved to scheduling a comprehensive visit to coincide with a patient's birthday visit. During this, the site includes emergency care planning and orders a TCD.

Addressing Psychosocial Needs

Holistic care of the patient includes addressing the mind, body, and spirit. Children living with SCD are reported to have poorer psychological and social well-being compared to demographically similar children living without SCD (Palermo et al., 2002). SCD affects the psychosocial health of parents and caregivers of children with SCD, including worrying more about their child's health, depressive symptoms, and internalized stigma (National Academies of Sciences Engineering and Medicine, 2020). Further, people with SCD also are at risk for cognitive deficits resulting from stroke or anemia, which can impact adherence to treatment (National Academies of Sciences Engineering and Medicine, 2020; Prussien et al., 2019).

The myriad of psychosocial needs were an important area of concern among program sites, and sites used both traditional care and innovative programming to address psychosocial needs. Appendix A: RCC Activities has additional examples. The Program developed a recommendation on the need to be responsive to psychosocial needs.

Coordinated Care

People with SCD require several levels of care that often involve multiple specialists and can require several procedures (e.g., eye and spleen functioning exams, TCD, immunizations, bloodwork). Separate, serial appointments are taxing on both people with SCD (transportation, cost, time away from work or school) and providers (administration of scheduling multiple appointments, as well as the potential that each appointment creates the possibility of a missed appointment, which adds burden to healthcare

systems and uses limited resources). Therefore, many sites have prioritized establishing dedicated clinic days and space; coordination of care; and developing protocols for clear and consistent communication.

RCCs emphasized the particular importance of access to physical space, lacking at many sites. Dedicated clinical space to serve people with SCD offers benefits for both providers and patients, helping to foster consistent care at important times, such as the transition from pediatric to adult care. In addition, the co-location of providers, services, and population with the same condition helps ensure that tailored processes meet the needs of both the clinicians and patients. The Program offers a recommendation to support coordinated services. RCCs report that when care is coordinated, they are able to improve access and quality that, in turn, improves outcomes. Additional information about this topic and examples of care coordination can be found in Appendix A: RCC Activities.

Continuous Care

Maintaining communication and engagement with a patient and their family during times of routine care as well as critical times, such as care transition (pediatric to adult) are both essential to ensure quality care.

Telehealth and **telemedicine** have been an important option for maintaining routine health care for people who live in rural areas or have other clinic access issues – and virtual visits have been a tool for SCD patients, especially to address many of the barriers related to missed appointments. COVID-19 prompted clinical reforms out of necessity, the greatest being more regular use of telehealth. A rapid and significant reliance on telehealth and telemedicine was the most prominent way that SCD providers, patient care, and provider-patient relationships changed to meet the shifting landscape.

MAINTAINING PATIENT CONTACT

Telehealth is the use of two-way telecommunications technologies to provide clinical healthcare through a variety of remote methods and includes a broader scope of remote health care services, including non-clinical services.

Telemedicine is focused and refers specifically to remote clinical services. (American Academy of Family Physicians, n.d.).

Considerations

While the benefit and need for this method of providing care in 2020 and 2021 is unquestionable, implementation in the Program exposed issues, including technical and user challenges; variable internet stability for providers and patients; and patient's lack of technology or capacity to use video, which may be essential for clinicians to complete some aspects of a visit. And those with technological issues rarely had immediate access to case managers or other technical support services that could help them resolve issues to complete a visit. To address this problem, some RCCs are now planning to train community health workers to help patients with access through their CBO partnerships.

Many providers also worried about the length of time between labs or missed in-person clinical tests, such as TCDs, with a reliance on telehealth. Clinicians reported becoming comfortable adjusting pain medication via telemedicine, but modifying select treatments like HU requires in-person procedures such as blood tests. And finally, housing and economic circumstances vary; not all patients have privacy during calls and those limited by a pay-per-minute mobile phone plan may not be able to prioritize the minutes for a virtual health visit. Additional information is in Appendix B, and the Program developed a recommendation regarding telehealth and telemedicine access.

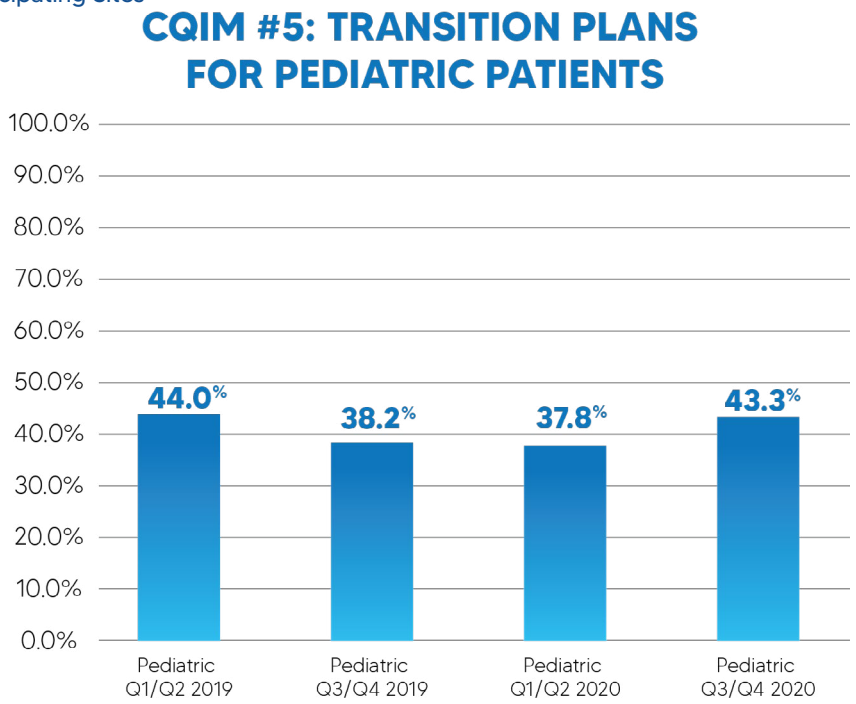
Bridging the Gap During Transition from Pediatric to Adult SCD Care

The transition period between pediatric and adult SCD care is particularly precarious; multiple studies have shown increased morbidity and mortality during this time (Darbari et al., 2019). A quality transition plan to prepare pediatric patients to receive care in healthcare systems focused on adults remains an important challenge and was a major focus for the RCC teams' work and a recommendation from the Program. Many participating sites are developing, testing, or have launched innovative processes to increase and improve transition care planning and programming. Some of the work has addressed better tracking of this healthcare component. This is vital as findings have shown that transition is not routinely and broadly tracked, making it difficult to estimate the percentage of pediatric patients who make this transition successfully (National Academies of Sciences Engineering and Medicine, 2020).

A critical first step in transition is to develop a **transition plan**, which is a documented, shared record to support planning for care as an adult. During the Program, several RCCs focused on improving the number and percentage of their pediatric patients with a transition plan. The teams felt they "owe it to patients to make sure they have a good setup when they leave pediatric care." Examples of the activities and strategies that RCCs used to improve transfer of care safety are in Appendix A.

Three specific studies show the need to increase transition programming as well as to rountinely track transition care. Two studies with pediatric patients with SCD (Andemariam et al., 2014; Hill et al., 2014) estimate that 39 to 68 percent of pediatric patients successfully transition to adult care and one study of children with special health care needs overall showed only 21.6% of youth making a successful transition to adult health care (Got Transition, 2020; Oswald et al., 2013). Program sites reporting on this measure showed results relatively consistent with these studies. Approximately 40 percent of the sites reported transitions from pediatric care to adult care.

Table 14. Percentage of Pediatric Patients with Documented Pediatric-to-Adult Care Transition Education, Aggregated into Six-Month Increments Among RCCs and Participating Sites



There are clear challenges when a person living with SCD is age-eligible to be transitioned from pediatric to adult care. Currently, there is a significant shortage of qualified providers to take care of the SCD population, especially qualified adult care providers, and a need to establish and support an ongoing pipeline of adult care providers. Additionally, not all healthcare systems have established transition programs, so the process to move a patient may not happen quickly or smoothly. Finally, Medicaid may end or be interrupted when a young adult turns 19, compounding the problem of ensuring uninterrupted care into early adulthood. Additional information is in Appendix B and in the Program recommendation for this area.

An Essential Element for Quality SCD Care: Engaging and Partnering with Patients, Families, Caregivers, and Community-Based Organizations

Engaging patients, families, and caregivers in their own medical care has resulted in improved adherence to medication, better self-care, and increased patient satisfaction. Additionally, CBOs have an important role in helping patients and their caregivers engage in their own care, and to serve as partners with clinics, providers, patients, and families who seek to learn more and improve health outcomes.

Engaging Patients and Families to Increase Accessibility of Appointments and Remove Barriers to Care

Missed appointments create financial and administrative issues and, importantly, gaps in care that may or may not be able to be rectified over time. Tracking and monitoring important clinical aspects, such as medication dosage, and meeting the patients' broader needs is difficult. Program sites reported the need to improve patient engagement to achieve better appointment and healthcare completion.

Transportation

RCCs and their participating sites repeatedly expressed concern about transportation issues. Unreliable transportation plagued patients in both rural and urban locations but was particularly felt in remote locations where providers recounted patients driving up to six hours for an appointment or to pick up a prescription. Some RCCs have set up satellite clinics and are expanding outreach efforts to address this barrier.

Insurance

While complications or lack of insurance keeps patients from accessing medical appointments, providers in the Program shed light on an area that has been equally consequential: the challenge for some patients to access life-saving medications. This report notes the support for prescribing HU and other proven and emerging medications to reduce symptoms and improve quality of life for people with SCD. Yet not all formulations of HU are covered by insurance and emerging therapies are not covered by all insurance plans.

Patient-Centered Information

Quality care includes targeted and accessible patient education materials, which removes barriers related to literacy, language, and culturally appropriateness. The RCCs developed materials for both providers and patients. Additional discussion is in Appendix B, and materials and resources are in the Compendium of Tools and Resources. Additional information about reducing barriers to appointment attendance and examples of RCC work in removing appointment barriers are in Appendix B.

Partnering with SCD Community-Based Organizations

A partnership between clinics and CBOs strengthens quality care for people with SCD; the Program focused on increasing this important collaboration. CBO involvement is essential to move SCD care forward, as they are instrumental in building relationships with patients, caregivers, and the community, and helping educate providers. As one CBO leader put it, "Our first mission is to maximize the quality of life of individuals living with sickle cell disease." Each of the RCCs have cultivated strong relationships that authentically engage the community, whether through expanded partnerships with CBOs, creating and supporting programming, or developing other ways to reach the population. The Program has made a recommendation for continued partnership.

Those who lead community efforts are passionate about this work and often have deep-seated community roots. The strength of the CBO partnerships with clinics comes from the longevity and constancy of these leaders' commitment to and understanding of the community the serve. CBOs who partnered with RCCs increased the value of the RCC, and all RCCs reported that partnering with CBOs enhanced the effectiveness of their work. CBOs with stronger infrastructure, established staffing, and funding sources outside of the Program accomplished more. The CBOs who participate with the Program spoke of the challenges they faced as they built their programming to current levels and welcomed ongoing work with RCCs to continue the partnerships that have been forged during this funding.

Throughout the Program, CBOs worked with RCCs in a variety of ways:

Pacific

- One CBO has run two long-time camp programs: Camp Gibbous for teens and Camp Crescent Moon for younger children. These programs provide summer experiences for the campers and for teens and young adults living with SCD working as counselors and in other roles.

Heartland/Southwest

- The CBO in St. Louis has been a funded partner with the Heartland/Southwest RCC for the past two funding cycles. They regularly host community education and awareness events, including provider panels and discussions on the importance of participation in research; this CBO also has championed the development of task forces to improve access to quality care for people living with SCD.

Midwest

- One site invites parents whose babies have been diagnosed with SCD to a series of educational sessions that allow participants to interact and find a system of peer support in addition to the important information.

Northeast

- The lead CBO convenes all local CBO leaders monthly to collaborate and work collectively. Supported by this Program, the lead CBO funds smaller CBOs to conduct activities across the region.

Southeast

- One CBO sets regular monthly meetings for patients, where they can share concerns and needs. They sponsor events to raise funds for scholarships for high school seniors who have SCD.

More information about CBO partnerships and examples of CBO activities are in the Appendix.

Lessons Learned, Recommendations, and Final Thoughts

The Program collected data focused on goals and objectives intended to improve the health of people with SCD. Collecting data from health systems is challenging and therefore the Program lessons learned relate to data collection.

Lesson 1: Capitalizing on Prior Work

To capitalize on prior program work, data measures should be consistent over time. Consistency will allow sites to continue established systems and improve ones that need editing instead of restarting new efforts each funding period.

Lesson 2: Value of Qualitative Data Collection

While a formal qualitative interview process was not originally planned as part of this project, pivoting from general site visits to individual in-depth interviews to accommodate COVID-19 restrictions provided an opportunity to gather rich experiences directly from SCD providers across the country. Future data collection would profit from continuing qualitative data collection and adding both site visits and individual interviews to obtain the most complete representation of regional and participating site work.

Lesson 3: Piloting a Provider Survey

Having a national perspective of providers who care for people with SCD is important. Collecting the PSPM data at two timepoints (2019 and 2020) was important in reflecting provider attitudes, areas of specialty, and types of care provided. However, to be able to compare survey answers across time points, having a uniform definition of the group who should be sent the survey ahead of survey fielding is necessary. While both survey results provided informative data, the results of the second provider survey – where a uniform definition was employed – will be useful to build upon in future iterations of the Program.

Lesson 4: Consideration of Differences in State and Local Variances

Public health guidelines and resources, as well as variations in size, experiences, and resources of the participating sites, impacted RCCs' ability to collect data. An assessment of these types of variations will be helpful before implementing the next data collection.

The work needed to ensure high-quality, consistent data collection can require significant effort and expense, which is important to consider. As the next iteration of the Program is planned, the above lessons learned may be useful when creating the measure sets and data dictionaries.

This report contains a complete set of recommendations in clinical care, healthcare policy, and ongoing programming and future initiatives regarding SCD care.

The 2017-2021 Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program addressed clinical and psychosocial needs to improve the health and quality of life of people with sickle cell disease. The Regional Coordinating Centers conducted numerous activities in the Program priority areas, particularly the three healthcare domains described in this report. These activities together with the Program recommendations will inform and enhance future efforts to provide quality care for people living with this complex condition.

Appendices and other documents provide additional information about subjects covered in this Report to Congress, including SCD and its impact on those with the condition, their families, and caregivers; methodology and data collected for the Program; a compilation of select RCC activities; summary of the impact of the COVID-19 Pandemic on the efforts of this Program; and learnings from the Program, all of which can be found in the Appendix and the Recommendations. Additionally, this report includes a Model Protocol and Compendium of Tools and Resources readily available for use by any organization or healthcare system seeking to improve the health and lives of people living with sickle cell disease.

Impact of the COVID-19 Pandemic on the Program

The U.S. Department of Health and Human Services declared a public health emergency on January 27, 2020, because of confirmed cases of 2019 Novel Coronavirus. COVID-19 caused significant disruption to health care systems throughout the U.S., and many SCD providers in the SCDTDRCP were engaged in either front line care or planning and developing procedures and processes, or both, to respond to evolving needs. Healthcare systems and individual providers quickly implemented new workflows and treatment approaches to provide essential care for people living with SCD -- who were at high risk for serious complications of COVID-19.

The pandemic directly affected SCD care and the implementation of the Program for at least 18 months, half the Program. Across the board, RCCs had to temporarily halt or change their operations, which altered some clinical and CBO services for people with SCD. These changes impacted the activities and data collection of the Program.

SCD Clinical Care and CBO Services

Though clinics and care teams could not operate under standard procedures they pivoted to support their SCD population and their organizations as well as they could. See Appendix A for RCC COVID-related activities. Given the potential of deadly infection for this immunocompromised patient group, clinicians did everything they could to keep their patients out of EDs to reduce the possibility of virus infection.

While there was variation by state, clinic and location, programs implemented strict changes to their clinical operations and community offerings, including:

- Staffing triage calls 24 hours a day so no patient would go to the hospital before speaking to someone on the SCD team
- Allowing only one caregiver in pediatric practices, and no companions in adult care units, except to support patients with cognitive dysfunction
- Providing protection for adult patients with end organ damage, especially lung disease, to bypass waiting areas and go directly to patient care rooms
- Working with hospital administration to give up dedicated SCD day-clinic space such as for blood transfusions to in-patient rooms for those sick with COVID-19
- Merging pediatric care centers to allow for opening more adult care locations, given the number of adults needed treatment for COVID-19

As COVID-19 halted in-person meetings across the U.S., the ability to regularly share information among providers in person decreased and, in many cases, abruptly ceased, as did most community programming — having to be canceled or modified to a virtual platform. RCCs found that while they lost the opportunity for informal teaching and sharing, the established structure of the Program network allowed them to continue their ability to quickly exchange information through telementoring such as Project ECHO® which became essential during the pandemic. RCCs capitalized on their experience using this model to expand COVID-19 offerings to quickly meet the information and education needs of attendees.

COVID-19 prompted the necessity to offer telehealth for patient/provider medical visits more broadly and frequently. While some clinics had deep experience providing direct-to-patient telehealth prior to COVID-19, it was not highly utilized in many clinics, resulting in implementation challenges and barriers (such as equipment or device access, connectivity, technology literacy, inability of some patients to use video visits, etc. for both patients and providers). Given that telehealth is likely to continue at higher levels than pre-pandemic, these issues must continue to be resolved. Further, while telehealth appointments are a valuable option, they cannot replace all clinical appointments for this population; some services must be completed in person (for example, TCDs, medication adjustment, acute pain management, transfusions, assessing disease progression). Positively, reimbursement mechanisms were quickly established to support this important care offering and RCCs recommend that such mechanisms continue for care continuity.

Program Data Collection Impact

A core activity of the Program was data collection to measure select aspects of SCD care. The following describes how the three data methodologies were impacted by the pandemic.

Effects of COVID-19 on the PSPM Data Collection

Originally, three annual PSPM surveys were planned. The second survey was originally slated to begin May 1, 2020. By April 2020, it was clear that fielding this provider survey was not a reasonable ask of RCCs or potential respondents. The Program decided to delay the May 2020 survey and launch it in September 2020. Due to timing, the September 2020 PSPM was the final survey of the Program. Therefore, the Program fielded a total of two provider surveys (vs. a planned three rounds of fielding).

Impact of COVID-19 on Clinical Quality Improvement Measure Data Collection

In March and April 2020, the NCC held conversations with RCC teams during individual monthly check-in calls to learn how the pandemic was impacting programs, local sites, and capacity to collect data; all sites believed they could collect and submit CQIM data as scheduled. The RCCs were in close contact with most sites to monitor data submission progress; for example, in Q1 2020, furloughs in two regions meant that some sites were unable to submit data. The RCCs annotated in NICHQ's CoLab and the NCC noted the fluctuation of sites submitting data during the course of the Program.

Qualitative data were collected for a comprehensive picture of the activities of the Program. Originally, the NCC planned to attend regional meetings in person to collect information about RCC programming and activities from all participants. Due to the pandemic, these data were limited and collected through virtual interviews with all RCC leads and up to three sites that were identified by each of the RCCs.

While the preceding describes aspects of the known impact of the pandemic on data collection, the RCCs relayed other ways in which COVID-19 may have impacted reported data. For example, to avoid the risk of exposure at large health care systems, immunization may have increased at community locations such as schools and community pharmacies, where documenting completion of this service might be missed or not included in a person's health record. As noted in the data appendix, this could have exacerbated fractured record keeping and unanticipated reduction in reporting. As well, several other factors could have contributed to areas of data collection, such as patient reduction in completion of in-clinic appointments either because of personal fear, clinical capacity or decreased access to transportation. This could have reduced numbers of measures completed (i.e., TCD, immunizations, HU prescriptions). Finally, due to other clinical demands, QI projects stopped or slowed during the course of the pandemic potentially impacting progress in some areas.

For more information on how the pandemic affected the Program data collection efforts, see Appendix C: Data Methodology. For more information on the ways RCCs pivoted to ensure critical services during the COVID-19 pandemic, see Appendix A: RCC Activities.

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SICKLE CELL DISEASE TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Recommendations

REPORT TO CONGRESS
SEPTEMBER 2021

Recommendations

Introduction

This section documents recommendations for achieving high-quality, comprehensive sickle cell disease (SCD) care. A broad range of stakeholders, including RCCs, Community Based Organizations (CBOs), Oversight Steering Committee (OSC) experts, and the National Coordinating Center (NCC), developed these recommendations. Some recommendations are based directly on work from the Program. Other recommendations come from general clinical and patient care experience. Links to recommendations are found throughout the Congressional Report.

Recommendation Categories

Three central themes emerged:

1. People living with SCD need to receive the best possible **clinical care** if they are to experience optimal health and well-being. **Clinical Care Recommendations** primarily address direct services. These can be improved and expanded in many ways, including more use of local and national registries; education campaigns for both patients (e.g., up-to-date information about standard and novel therapies) and providers (e.g., pain management, essential transition care elements, guideline-based care); and designing and using systems that monitor key processes and outcomes, such as immunization, TCD screening, and transition of care.
2. **Healthcare Policy** plays a central role in supporting the diverse and complex needs of people living with SCD. **Healthcare Policy Recommendations** support coordination of services, access to care, ensuring there is a plan to provide the full array of psychosocial services needed, and supporting a pipeline of adequately trained SCD providers.
3. Through the Program work, **ongoing programming and future initiatives** were identified. **Future Initiatives and Programming Recommendations** address programmatic structural recommendations, which could be applied now or in the future. Topics include organizing national efforts; choosing which types of organizations should be hub centers; and the benefit of using registries.

Foundation Tenets

In addition to the specific recommendations provided, three key foundational areas were identified as critical to the success of current and future programs. Comprehensive SCD care requires:

1. Measurement and Assessment to Support Continuous Improvement

In all areas, continuous measurement, evaluation, and the expectation to be responsive to findings are necessary to ensure that high-quality care is being provided. A common set of standardized measures and a framework to guide improvement work will continue to be critically important. Quantitative and qualitative data collection during the Program was essential for teams to identify successful strategies and best practices. In addition, having a reliable way to capture, organize, and share data was crucial. For each recommendation, the process and outcomes should be measured and shared from start to finish.

2. A Diverse Set of Stakeholders

Efforts to improve care for people living with SCD must include: clinical staff (specialty and primary care) from large and small practices and centers of excellence; support from behavioral health and social services; CBOs; administrators who can help manage and spearhead registry efforts; and people living with SCD and their family members. No single group can achieve success alone.

3. Resources and Support

Given the complexity of the disease and needs of families, resources are required for:

- Quality measurement, evaluation, and adjustment as needed, including reprogramming databases or increasing staffing availability
- Direct services, especially for care team members who specifically focus on addressing psychosocial and behavioral health support
- Provider training across the career span of clinicians
- Support and leveraging of CBOs who can offer an array of services to support people with SCD and enhance the ability of clinical care teams to be responsive to patients' needs
- Maintaining and updating national registries

Recommendations Emerging From the Program Work

Clinical Care

1. Leverage existing registry information and improve current EHR systems (e.g., EHR templates; order sets; tracking and feedback mechanisms).

- Data from these sources can be used to improve the care of patients with SCD as well as provide a national perspective of the patient population.

- The use of available registries, EHR templates, order sets, etc. should continue. The experience through the Program showed that there were varied outcomes in the use of these types of resources, but continuing work to improve all is recommended.

2. Monitor key processes and outcomes with a focus on achieving the high-quality, comprehensive care recommended by national guidelines, including screening and preventive interventions (e.g., penicillin prophylaxis, immunizations, HU use, transfusion, TCD screening) as well as ED visits, hospitalizations, and readmissions.

- Consideration should be given to stratifying key process and outcome measures by race/ethnicity and other relevant social determinants of health (e.g., income level, zip code, language) to assess for within and across group disparities.

- RCCs followed national guidelines; clinics that had buy-in and infrastructure to support use of these guidelines are models to replicate.

- Data tracking the use of select national guidelines was completed and are included in this report. This type of tracking requires significant staffing and resources. Disaggregation of data is important to better understand details of how care is being provided but was not in the scope of this program.

3. Expand the use of evidence-based care plans and other care coordination tools for individuals with SCD.

- Measure this process, as it will inform how to best use and optimize plans.

- Emergency care and related pain management protocols and strategies were not core areas of measurement under this funding. However, several sites have prioritized these topics. Materials can be found in the Compendium of Tools and Resources and Model Protocol sections.

4. Provide education regarding use of therapies (i.e., HU, transfusion, other disease-modifying therapies) for individuals living with SCD and their families.

- Using evidence-based shared decision-making tools that support discussions of benefits and risks, as well as patient preferences and strategies for self-management support, is beneficial.

- Clinics and CBOs completed patient education across a broad spectrum of topics. Several are included in the Compendium of Tools section.

5. Provide supports needed so that healthcare systems are equipped to provide robust care coordination for both psychosocial and medical needs of individuals living with SCD and their families.

- Healthcare teams for both the pediatric and adult populations should include social workers, mental health specialists, community health workers, occupational therapists, and similar providers to address needs, reduce fragmented care, and ensure bias is minimized and health equity maximized.

- ECHO sessions could be dedicated to each of these specialty areas for greater reach.

- The staff time needed to develop protocols and oversee implementation must be considered.

- Psychosocial areas are longstanding needs for patients with SCD and work through the Program has again confirmed this. But the critical role of mental health emerged as an under-supported and urgent priority, especially during the pandemic.

- The time needed to fully develop and implement a thoughtful and cohesive plan for all healthcare systems was not in the scope of this program but should continue to be priority moving forward.

6. Provide supports to ensure that all facilities providing care for individuals living with SCD incorporate the following six core elements of transition where appropriate:

1. Having a transition policy
2. Developing a process for tracking and monitoring transition-age youth
3. Assessing and using transition readiness assessments
4. Planning for transition
5. Transferring care
6. Completing transfers

Some sites have mastered all six essential areas; others need to address some, while others must begin. Supporting teams as they work to achieve mastery across the board for strong transitional care is welcome.

7. Include individuals with SCD and families in both the design and implementation of efforts to improve care.

- Reference and incorporate existing program/initiatives as applicable (e.g., Foundation for Women and Girls with Blood Disorders; American Academy of Pediatrics Learning Action Network efforts).
- All providers working with individuals living with SCD understand and support the emphasis on and need to engage families. Cross-learning continues to be important and using already available resources designed for this effort is worthwhile and efficient.

8. Develop and disseminate standard models and/or curriculum to improve knowledge related to SCD care for all members of the care team.

- Care team should include physicians (specialists and generalists) and advanced practice providers (nurse practitioners, physician assistants, nurses, social workers, community health outreach workers, mental health specialists, physical therapists, etc.)
- Training should begin early (undergraduate medical education and post-graduate medical education) and be an ongoing requirement for continuing medical accreditation
- Education through ECHO, other medical direct education opportunities, and conferences continued during the Program. Clinics, CBOs, and individual providers tried various routes to support improved knowledge and experienced different levels of success.
- While developing detailed standard models/curriculum related to SCD care was outside the scope of this program, the Program supports ongoing work in this area.

Healthcare Policy

1. Support expanded access to evidence-based SCD care through telemedicine, telehealth, telementoring, and other innovative models for these services.

- All RCCs and some of their local sites have supported, participated in, and implemented telemedicine, telehealth, telementoring, and other innovative models for these services. These types of services became particularly important given the special needs during the pandemic. Given the duration and success of these components, the Program anticipates that continuation and expansion going forward will be important.

2. Advocate and promote improved access to digital technology and internet access for people living with SCD. This is particularly timely given the needs of people living with SCD and their families who may face barriers to accessing in-person care (e.g., due to personal circumstances or unforeseen events, such as COVID-19).

- Telemedicine, telehealth, telementoring, and other innovative models require the use of technology, both by providers and patients. Some clinicians experienced variability in their patients' ability to participate in all electronic methods. For instance, some patients relied on pay-by-minute phones and could not expend valuable minutes for a phone visit, while others did not have video capability.
- Continuing to improve access to digital technology and internet access will be important in the near and distant future.

3. Advocate for sustained and enhanced reimbursement for clinicians and complex care teams providing traditional as well as telehealth care. SCD should be automatically added as an eligible condition for enhanced reimbursement.

- Through the work RCCs conducted during the Program, they saw the need for sustained and enhanced reimbursement for team-based care. However, pursuing change regarding reimbursement eligibility criteria was out of the scope of this program.

4. To strengthen quality SCD care, incorporate standardized quality measures of access and care for individuals living with SCD into organizational performance measures, such as the Bureau of Primary Healthcare quality metrics and the new American Society of Hematology guidelines for care. These measures should be tracked in existing registries.

- All providers should be encouraged and supported to employ quality metrics that align with the Centers for Medicaid and Medicare Services Equity Plan for Improving Quality in Medicare to maximize use of these standardized measures. Adherence should be incentivized to encourage uptake.
- Harmonization and standardization of quality measures and metrics, such as put forth by trusted agencies such as the American Society of Hematology, is essential to ensure consistent, national care.
- At this time, work from the Program shows that RCCs and their sites have a strong sense of the guidelines, but greater consistency is needed.
- Work should continue on this recommendation in future program iterations.

5. Create system infrastructure and adequate reimbursement for care transition throughout a patient's lifespan that ensures seamless and comprehensive care.

- Strong team-based care should be employed and include a variety of providers. Care of patients living with SCD should involve both primary care and specialty clinicians. When available, specialists should be encouraged to lead and coordinate care in collaboration with their primary care colleagues. The SCD expert, medicine/pediatrics internists, and/or family practice providers should work together to meet the medical needs of the patient.
- At all times, but especially at crucial transition points, support care team members, such as social workers, care coordinators, and mental health clinicians, must be an active part of the care team.
- During the Program, many RCCs and their local clinics were focused on transitional care and grew the implementation of quality care in this area. However, data from the Program showed continued attention is needed. Please see the Compendium/Model Protocol for specific resources.
- In addition to the focus on transitional periods of care, there is great interest and support for implementing effective care across the entire lifespan of patients with SCD as noted in this recommendation. The experience of the Program teams was that coordination of care is best led by a provider who specializes in SCD care, someone who deeply understands the disease and essential care components that must be tracked, addressed and continued through completion. In particular, providers at or affiliated with a comprehensive SCD center bring important experience and are preferred for this role when possible.
- Overall, additional structure and support is needed to address infrastructure and reimbursement barriers, which was not in the scope of this funding.

6. Support tailored training of qualified healthcare professionals who understand collaborative care models and complex care for both pediatric and adult patients with SCD.

- Students have different learning needs from licensed practitioners. And, within providers, attention should be given to determine whether APPs (NPs/PAs) need a different educational track from physicians. Exposure to evidence-based content to build knowledge and skill may be the same, but intervention strategies may be different.
- Building a pipeline of providers willing to specialize in SCD care will be important to ensure high-quality care for this HRSA-designated medically underserved population. Student loan forgiveness programs for broader categories of physicians (e.g., hematologists), nurses, nurse practitioners, and physician assistants should be considered.
- Throughout the Program, training was important and it is recommended that this be a core part of future work. Grand rounds, SCD-specific training (including biological pathways, treatments, and psychosocial needs), ECHO®-based learning, and other educational opportunities must continue. Having a national site that has links to all recordings of program ECHO® sessions would be beneficial for shared learning.
- While continuing medical training opportunities must continue, a more focused plan is required to increase the number of providers specifically trained to take care of patients living with SCD. Such a plan should include making it more financially appealing. This work fell outside of the scope of this program.

Future Initiatives and Programs

1. Continue to support a regional infrastructure for the Program.

- This approach worked well and should be maintained since it supported strong delivery of care across participating sites.
- The regional infrastructure should continue to encourage funding that fosters and supports all clinics providing care for people with SCD. Variety of size and geographic location of clinics is recommended. Collaboration with CBOs and community-based practices to enhance delivery of care is recommended.

- Within a regional infrastructure, RCCs previously used the Collective Impact Model (CIM) effectively. The CIM promotes using common agendas, communications, data, and mutually reinforcing activities, and supports the role of the RCCs as backbone agencies of the regional structure. Resuming a CIM framework within a regional model allows for building the synergies necessary for building capacity. Reinstating the use of CIM should be considered.

- A regional approach was adopted by all regions. Further, reflecting on the breadth and depth of work accomplished during the Program, it was seen to be a cost-effective approach. The Program also recommends when using this approach, greater site/clinical/CBO coverage and appropriate resources must be provided to experience the most powerful impact, as financial limitations hampered increasing the reach, benefits, and power of a true learning collaborative.

2. Support the growth of the following monitoring and awareness areas via funding and coordination.

- Utilize the national SCD registries that already exist. Per the 2020 National Academies of Sciences, Engineering, Medicine report *Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action*, use of a national, ongoing clinical registry in conjunction with a surveillance program in order to enhance quality care in SCD is recommended.

- Increase SCT education and counseling re: inherited genetic risks.

- Coordinate national SCD surveillance with the CDC Division of Blood Disorders and other federally funded programs to maximize benefit.

- Data was a major focus of this program and RCCs and their local collected the measures reflected in the data section, but collection methods could be strengthened.

- Surveillance is needed to better measure quality and there needs to be a robust, efficient process to support this.

- All RCCs support use of registries, but capitalizing on current ones and improving coordination of these resources was not within the scope of this program.

3. Continue support for and expand the Project ECHO® model by developing standard SCD curricula and metrics of success. A primary objective of future programs should be to increase the number of providers who are knowledgeable about treating SCD.

- In addition to ECHO-based work, support telehealth efforts which have been shown to improve access to services for persons with SCD, especially those who have had difficulty accessing care previously.

- All RCCs and some of their local sites, have supported, participated in, and successfully implemented ECHO-based learning.

- RCCs would like to continue to implement and expand ECHO programs if feasible.

- Developing standard SCD curricula will be important to address in a future funding iteration, as it was not in the scope of the current program.

General Recommendations to Support Improved Care for Individuals with SCD

Clinical Care

1. Address deficiencies in pain care during ED and other visits for persons living with SCD by establishing and widely disseminating:

- Tailored pain management plans;
- Institutional pain management protocols; and
- Strategies to promote use of more easily administrable pain medications.

Emergency care and related pain management protocols and strategies were not core areas of measurement under this funding. However, several subgroups have prioritized these important topics and valuable work continues in these areas. Thoughtful materials can be found in the Compendium of Tools and Resources and Model Protocol sections.

2. Health systems/institutions should universally screen individuals at risk for SCD and SCT (including immigrants and refugees, if not previously screened in their home countries) and link identified individuals to systems of care.

- The profile of people living with SCD in the United States is expanding. Individuals with SCD are arriving who may not be identified by newborn screening in their country of origin. Greater attention must be taken to ensure that these individuals with disease are offered appropriate care. This starts with culturally and linguistically appropriate education about SCD and screening for the condition. While it was not in the scope of this program to fully address this recommendation, the Program encourages newborn screening programs and other applicable health care systems to address this emerging problem.
- The U.S. Department of Health and Human Services can be an important resource when working with an immigrant population. The Health and Human Service regional offices offer links to state refuge health coordinators, who may assist in improving access to services. A state coordinator list is available on the HHS regional website: https://www.acf.hhs.gov/office-of-refugee-resettlement.

3. Advocate for adequate funding for preventive clinical and social services for persons living with SCD.

- Through Program efforts to improve preventative care, teams encountered substantial barriers to accessing services (preventive and social) and reasonable reimbursement for those services.
- Work to address these issues was outside the scope of the Program, but these are concerns that should be addressed in the future.

Healthcare Policy

1. Support efforts to ensure that all people with SCD (pediatric and adult) have consistent health insurance so they can get uninterrupted care, including access to newly approved treatments.

- Covering all people with SCD with Medicaid should be considered given the state variability of disability approval. And care must be taken to write insurance coverage flexible enough to address emerging clinical findings (e.g., pre-cirrhosis liver injury in chronically transfused children, neuropathic pain, and disabling bone tissue death).
- This recommendation is a supplemental finding related to implementation of the work.
- Addressing gaps in health insurance is imperative to ensure quality, comprehensive, life-long care. While of utmost importance, work on this specific issue fell outside of the scope of this program.

2. Adjust payment policies and enhance reimbursement rates to cover care coordination services that include community healthcare workers to improve access to community resources (e.g., non-emergency services such as transportation, social services, mental healthcare, and clinical services) for all patients, but especially for Medicaid/Medicare recipients.

- This should be done by working with Centers for Medicare and Medicaid Services to develop feasible payment systems.
- Support and psychosocial services for patients with SCD is needed. While mental health was not specifically addressed in the original FOA, this need gained prominence, particularly in the last year.
- Work is needed to adjust policies and enhance reimbursement rates, but was outside of the scope of this program.

Future Initiatives and Programs

1. Ensure that CBOs are included in the Program efforts and encourage them to serve as hubs.

- Once established, encourage CBOs to add additional spokes in each region that include clinicians and other CBOs.
- Prioritize sites caring for populations outside the current Program catchment areas throughout this process.
- During the Program, there was some variation in the relationships between CBOs and RCCs; some relationships were strong and some needed additional support.
- Movement to increase integration, especially to move CBOs into a hub role, will be important to address in future program iterations; this effort will be critical to improving work with patients and their families.
- Vast expansion was limited by funding availability.

2. Increase planning and communication between all federal SCD programs, especially the Program and SCD Newborn Screening programs, so that there is alignment of work timing as well as agreement about expectations of this collaborative effort.

- This will help ensure that both programs focus on providers and patients.
- Mutually reinforcing activities across both programs is essential to accelerating critically-needed improvements in SCD care. Improved synergy will support efforts to strengthen the linkages between individuals living with SCD, their families, and communities to services provided by CBOs and community health care workers.
- This structure is necessary for true coordination and to better understand SCD-related care nationally. However, the time, staffing, and infrastructure to do this was outside the scope of this funding period.

Glossary

accessible: Accessible means a person with a disability is afforded the opportunity to acquire the same information, engage in the same interactions, and enjoy the same services as a person without a disability in an equally effective and equally integrated manner, with substantially equivalent ease of use. The person with a disability must be able to obtain the information as fully, equally and independently as a person without a disability in terms of literacy level, language, and culture. Source

Acute Chest Syndrome: A condition affecting the lungs that is defined as a new radiodensity on chest radiograph accompanied by fever and/or respiratory symptoms. Damaged lungs lose their primary functionality and are a leading cause of death for people living with SCD.

Adakveo (crizanlizumab): A monoclonal antibody developed by Novartis that was approved by the FDA on November 15, 2019. Crizanlizumab, which is designed for people 16 years and older, helps reduce the frequency of vaso-occlusive pain episodes. This is important, as these episodes can escalate to life-threatening conditions and are a major cause of additional costs such as hospitalization.

advanced practice professionals (APPs): Medical providers (Physician Assistants and Advanced Practice Registered Nurses, such as Nurse Practitioners) who are trained and educated similarly to physicians. APPs are able to provide many of the same services as physicians (e.g., prescribing medicine)

allogeneic bone marrow transplantation: Cure for SCD in which a patient's own bone marrow is eliminated with chemotherapy and replaced with bone marrow from a donor

care transition: Process in which young adults with SCD transfer healthcare from pediatric providers to adult providers

Clustered Regularly Interspaced Short Palindromic Repeats (CRISPR): Experimental gene-editing cell therapy used in the treatment of SCD

community-based organizations (CBOs): Non-profit groups that help patients and their caregivers take part in their own care and serve as partners with clinics, providers, patients, and families to engage with all parties in order to improve health outcomes

electronic health record (EHR) systems: Systems of medical records on computers that enable tracking of clinical care elements (e.g., prescription rates)

Endari (l-glutamine): A drug designed for people aged 5+ that has been shown to reduce pain episodes requiring hospitalizations, as well as reduction in acute chest syndrome, compared to a placebo.

erythrocytapheresis: A non-surgical treatment red cell exchange transfusion

hematopoietic stem cell transplantation: The stem cells that form blood and immune cells are known as hematopoietic stem cells (HSCs). Bone marrow transplant, or hematopoietic stem cell transplant (HPSCT), involves the administration of healthy hematopoietic stem cells in patients with dysfunctional or depleted bone marrow. This helps to augment bone marrow function and allows, depending on the disease being treated, destruction of tumor cells with malignancy or generation of functional cells that can replace the dysfunctional ones in cases like immune deficiency syndromes, hemoglobinopathies, and other diseases. Hematopoietic stem cell transplants are now routinely used to treat patients with cancers and other disorders of the blood and immune systems. Source

Hib vaccination series: A series of vaccine doses to protect against Haemophilus influenzae type b (Hib)

hospitalists: Physicians who hold expertise in caring for patients in a hospital setting

hydroxyurea (HU, aka Siklos, Addmedica, Droxia): A medication that was not approved by the U.S. Food and Drug Administration (FDA) for adults living with SCD until 1998, and not for children until 2017. HU is a significant SCD therapy option as it has been shown to be very effective at reducing the frequency of pain crises and need for blood transfusions in pediatric patients aged 2 years and older who have SCD. Source

immunization: A prophylactic therapy that prevents life-threatening infections, with specific focus on pneumococcal vaccination

Institutional Review Board (IRB): Under FDA regulations, an IRB is an appropriately constituted group that has been formally designated to review and monitor biomedical research involving human subjects. In accordance with FDA regulations, an IRB has the authority to approve, require modifications to (to secure approval), or disapprove research. This group review serves an important role in the protection of the rights and welfare of human research subjects. Source

longitudinal SCD registry: This Registry was developed in Phase I and continues through Phase II with the goal of enrolling 2400 patients (300 per center) between the ages of 15 and 45 years to conduct comparative studies related to guidelines and recommendations and to address evidence-based management of SCD. It collects standard clinical measures, laboratory values, lifestyle factors, medical history, treatment, healthcare utilization, and patient-reported outcomes associated with pain, co-morbidities, quality of life, physical functioning, mental health, and barriers to care. Longitudinal data is collected on study subjects throughout the study period. The Registry is a resource for identifying gaps in research, conducting data queries and analyses that lead to development and implementation of research studies, and dissemination of research findings from the Registry data. Source

medical home: Where a practice-based care team takes collective responsibility for a patient's ongoing care (American Academy of Pediatrics)

National Coordinating Center (NCC): In partnership with HRSA, an organization that collaborated with the leads of the five regions to advance the common agenda of increasing access to high-quality comprehensive care that includes use of disease-modifying therapies for people affected with SCD. The NCC developed shared measures and the alignment (and improvement) of activities; enabled communication; and provided the overarching infrastructure, coordination, synthesis and dissemination of outputs, while also reporting progress to HRSA to ensure success. The 2018-2021 NCC was the National Institute for Children's Health Quality (NICHQ).

network: a nationally coordinated collaboration among and within five regions that shares understanding, commitment, priorities, clinical approaches, measurement, education, and community engagement strategies to improve the health of people living with Sickle Cell Disease (SCD) on a national scale

non-malignant hematology: A non-cancerous blood disorder. Sickle cell disease is a non-malignant (benign) blood disorder. Source Hematologists use the terms classical, nonmalignant, and benign hematology to reference the same entity: the study and management of nonmalignant disorders of the blood. It is a discipline unto itself, distinct from its sister field, malignant hematology. The scope of practice can be broad and includes thrombotic and hemorrhagic disorders, transfusion medicine, hemoglobin disorders including sickle cell disease and thalassemia, anemia, thrombocytopenia, leukocytosis, leukopenia, and disorders of iron metabolism. Source

Oversight Steering Committee (OSC): The OSC comprises experts who bring unique knowledge, skills, and connections that complement the knowledge and skills of the NCC team. The OSC serves to make recommendations and/or provide key information and materials to the NCC team and inform its decision-making. The roster can be found in Appendix B, Section 12.

pharmacokinetics-based guided dosing: Pharmacokinetics is the use of mathematics to determine the right dose for an individual.

pneumococcal disease: A bacterial infection that can affect the upper respiratory tract and can spread to the blood, lungs, middle ear, or nervous system

Project ECHO®: A guided-practice model that reduces health disparities in under-served and remote areas of the state, nation, and world. Through telementoring, the ECHO model uses a hub-and-spoke knowledge-sharing approach where expert teams lead virtual clinics, amplifying the capacity for providers to deliver best-in-practice care to the underserved in their own communities. Source

prophylactic therapy or treatment: A prophylactic is a medication or a treatment designed and used to prevent a disease from occurring.

prophylactic vaccine: A vaccine used to prevent a disease or infection

overt stroke: A type of stroke that results in apparent neurological deficits, such as weakness in an arm or speech problems Source

Oxbryta (voxelotor): A medication that can help reduce strokes by increasing hemoglobin levels. It was granted accelerated approval by the FDA just 10 days after submission, on November 25, 2019, for the treatment of SCD in patients 12 years of age+

quality-adjusted life expectancy (QALE): A model for clinical decision-making in which estimates of impairment or disability are factored into calculation of life expectancy. It is also a method of adjusting life expectancy to allow for reduced quality of life caused by chronic conditions. Many chronic conditions shorten life on average by predictable amounts. These expected amounts can be estimated from available sources, such as hospital discharge data and health survey data, and used to produce a QALE for individuals or subsets of the population. At the individual level, the QALE is based on clinical judgment and subjective opinions of patients about their quality of life, preferably arrived at by consensus between clinicians and patients. Source

Regional Coordinating Centers (RCCs): Regional Coordinating Centers (RCC) established regional networks and provided leadership and support for regional and statewide activities that developed and established systemic mechanisms to improve the prevention and treatment of Sickle Cell Disease.

sickle cell disease (SCD): one of the most common genetic conditions, caused by a single gene mutation that affects the red blood cells. People who have this mutation can experience a range of symptoms from mild to severe, and those symptoms can change during a lifetime with the disease. The mutation causes red blood cells to form into the shape of a sickle, with edges of the cells transforming from rounded to sharp. When the "sickled" red blood cells move through blood vessels, they can get stuck. The sharp edges pressing against the walls of the blood vessels can cause mild to severe pain episodes. In more severe cases, the sickled cells block the flow of blood through vessels, which results in significant illnesses.

silent stroke: A type of stroke that does not cause any noticeable symptoms and can only be seen on brain scans

telehealth: The use of two-way telecommunications technologies to provide clinical health care through a variety of remote methods

telemedicine: The practice of medicine using technology to deliver care at a distance. A physician in one location uses a telecommunications infrastructure to deliver care to a patient at a distant site. Source

Transcranial Doppler (TCD) screening: Transcranial Doppler (TCD) is a noninvasive ultrasound procedure that allows the clinician to clearly see how quickly blood is flowing through the brain over a period of time. High blood flow is associated with an increased risk of stroke. The test is reliable, painless, and relatively inexpensive.

transition plan: A documented, shared record to support planning for safe transfer from pediatric care to adult medicine

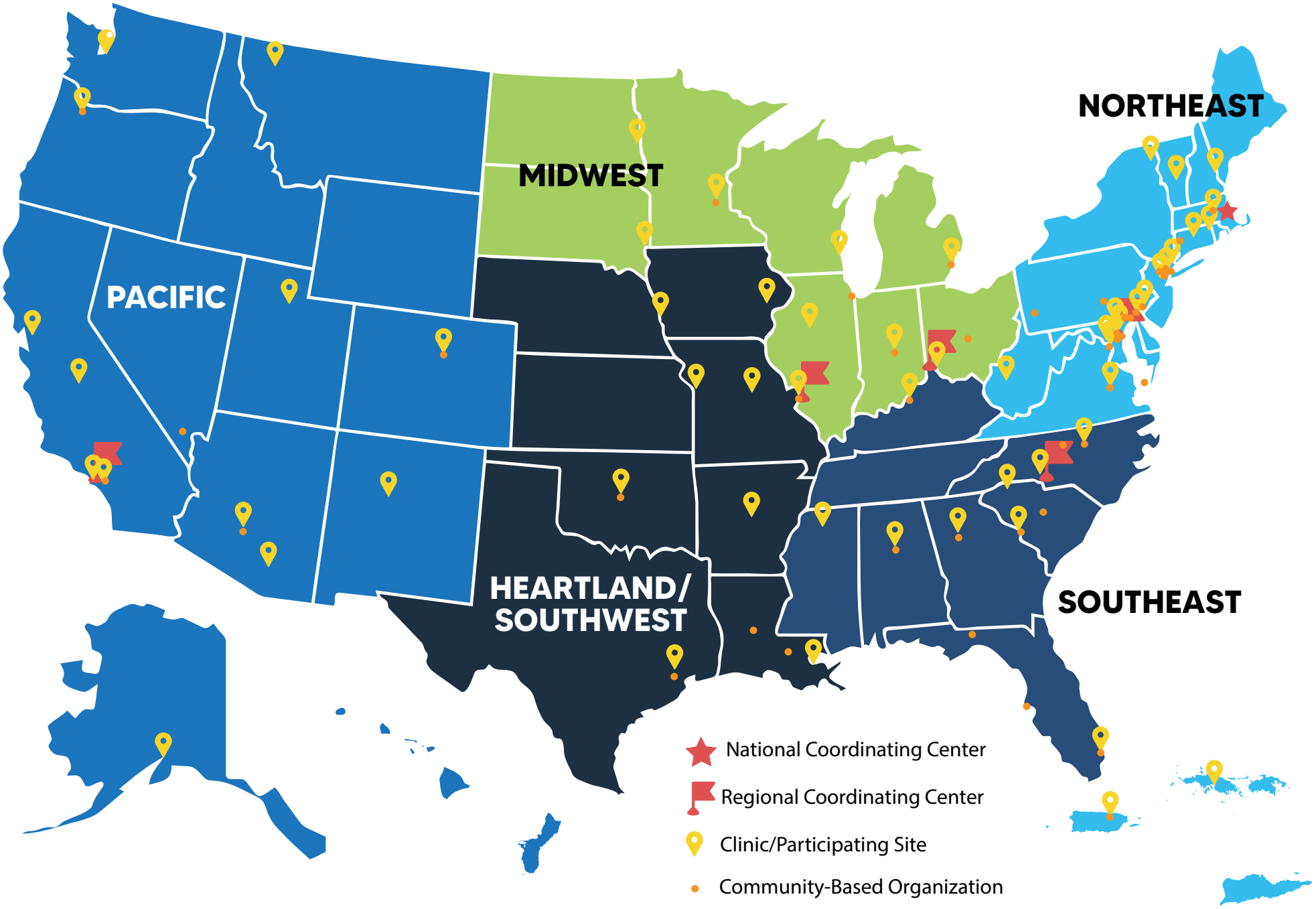
vaso-occlusive pain crises or episodes: Sickle cell anemia patients often experience episodes of acute pain that are caused by vaso-occlusive crisis (VOC). VOC is the most common complication of sickle cell anemia and a frequent reason for emergency department visits and hospitalization. Source

Appendix A: RCC Activities

REPORT TO CONGRESS
SEPTEMBER 2021

SICKLE CELL REGIONAL COLLABORATIVES

Figure 1. Map of National Coordinating Center, Regional Coordinating Centers, Clinics and Participating Sites, and Community-Based Organizations



PACIFIC REGIONAL COLLABORATIVE

Description of RCC Activities

Domain 1: Increase the number of providers treating individuals with sickle cell disease using the National Heart, Lung, and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report

Arizona

- Has created an electronic dashboard that tracks several clinical measures. A built-in algorithm helps identify and flag immunization and TCD due dates.

Colorado

- Has done innovative pain management work by spearheading the opening of an observation unit for acute pain management. This has resulted in a decrease of 20 percent in hospital admissions. This state also opened a community clinic in June 2020, which offers chronic pain management and accepts Medicaid coverage.

Nevada

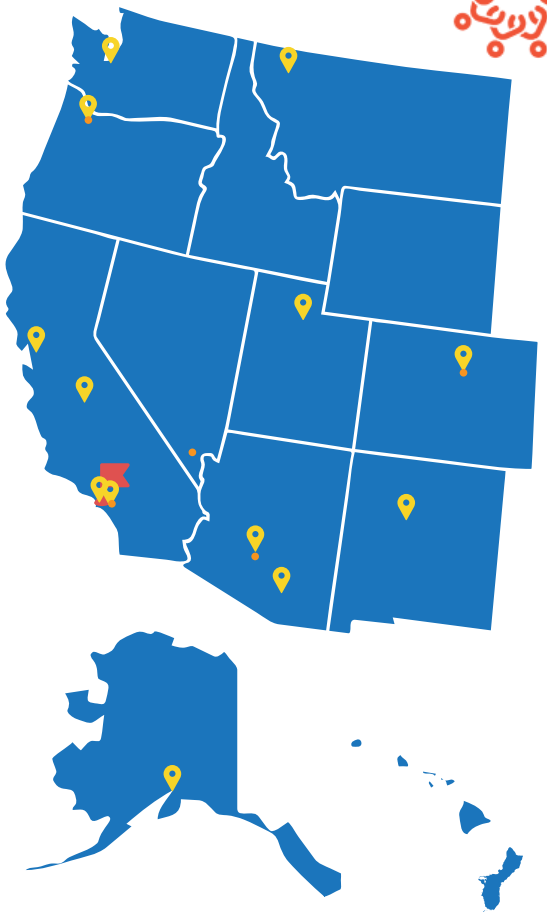
- Has conducted work in pain management. In addition to using the Oregon guidelines, this site has continued other work, including their H.U.G.S. (Holistic, Uncomplicated, Gratifying Support Services) program. This program is a holistic therapy program that focuses on pain management alternatives, such as meditation, massages, music therapy, etc. This site has also worked with the ED to try and reduce the amount of time people with SCD wait before receiving their first dose of pain medication.

Oregon

- Has leveraged their statewide database of vaccines that holds information for all patients born after 1990. This database allows clinicians to see all immunizations that a patient has received by date. To compliment this, the site has established a process for clinicians to look for what is due or needed and order these immunizations. This has been instrumental in helping ensure that patients receive needed vaccines at their next clinic visit.

About the Pacific RCC

PSCRC



Lead Organization

Center for Inherited Blood Disorders (CIBD)

MAP LEGEND

- Regional Coordinating Center (RCC)
- Clinic/Participating Site
- Community-Based Organization

Principal Investigators

- Diane J. Nugent, MD, Center for Inherited Blood Disorders
  Co-Principal Investigator
- Marsha Treadwell, PhD, UCSF Benioff Children's Hospital Oakland
  Co-Principal Investigator
- Elliott Vichinsky, MD, UCSF Benioff Children's Hospital Oakland
  Co-Principal Investigator

Data Managers

- Shalini Vora, MPH
- Priscilla Salceda, MPH

- Has conducted significant work in the pain management field. This site has implemented institution-wide SCD pain guidelines and tracking metrics. To create these metrics, it was important that the PI include multiple disciplines for clinic buy-in. She assembled 23 champions from around the hospital, including providers in anesthesia, pain, and inpatient/outpatient services. All team members came together to create this local pain treatment guideline. Oregon has shared these guidelines with others in the region.

Domain 2: Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care

State Plans

For the Program, RCCs developed a Regional Sickle Cell Action Plan and state-specific Sickle Cell Action Plans for funded states. The plans included 1) resources in each region and state to improve SCD care for all people with SCD in the region and 2) a description of the overall infrastructure that would address the goals and requirements listed in the FOA.

The state action plan described:

- How each state intended to develop a network of providers using evidence-based SCD care in the state
- How telemedicine/telehealth strategies and other provider support would be utilized
- How access to quality care would be supported
- How the state intended to increase the number of individuals with SCD being treated by providers using evidence-based SCD care

These plans were used to help formulate a cohesive approach to facilitate knowledge-sharing. Discussions spurred by these state plans brought forth a regional vision and strong partnerships with the RCCs that continues today. Information included may be helpful to states interested in doing similar work.

Pacific RCC State Plan

Participating Clinics

- AK – Alaska Pediatric Oncology
- AZ – University of Arizona Health Sciences Center; Phoenix Children's Hospital
- CA – Center for Inherited Blood Disorders; UCSF Benioff Children's Hospital, Oakland; Martin Luther King, Jr. Outpatient Center, Valley Children's Hospital
- CO – Colorado Sickle Cell Disease Treatment and Research Center, University of Colorado
- NM – University of New Mexico
- MT – Kalispell Regional Healthcare
- NV – Sickle Cell Center of Nevada
- OR – Oregon Health and Science University
- WA – Odessa Brown Children's Clinic, Seattle Children's Hospital
- UT – Utah Center for Bleeding and Clotting Disorders at Primary Children's Hospital

Participating Community-Based Organizations

- Sickle Cell Disease Foundation of California
- Bridging The Gap - Adult Sickle Cell Disease Foundation of Nevada
- Dreamsickle Kids Foundation (Nevada)
- Sickled Not Broken Foundation of NV
- Sickle Cell Foundation of Arizona
- Colorado Sickle Cell Association
- Sickle Cell Anemia Foundation of Oregon, Inc.

STATE PLAN LEVERAGE

In 2018, the Pacific region convened multiple stakeholders to begin the creation of a California State Action Plan. Using the SCDTDRCP FOA plan as a foundation, they sought funding from the California Department of Public Health to establish the Networking California for Sickle Cell Care Initiative. SCDF and CIBD shepherded the plan through the California State Legislature, obtaining $15 million in funding to improve care for adults throughout the state. Through CA's new legislation, infrastructure for a sustainable network of adult care was built.
Networking California for Sickle Cell Care

RCC ECHO Website

RCCs found the Project ECHO® approach to be effective and manageable for knowledge sharing. They reported high use and impact of Project ECHO® and expressed desire for continued support to expand this capacity.

Using this model increased RCCs' ability to offer SCD-specific regional and national sessions. RCCs were responsible for planning, organizing, and facilitating ECHO sessions tailored to their regional needs and convening national ECHOs as needed. Content covered SCD-focused information, from basic science to psychosocial needs. ECHOs often focused on region-specific content and cases to best support providers where they served patients. However, all RCC ECHOs were "open" and people from any geographic location within the country and world were able to attend. Some RCCs recorded the education sessions to ensure the greatest reach possible. RCCs noted that making ECHO open to all, live and recorded, was an important service for providers in their regions for whom they were not able to educate more directly due to geographic or resource limitations. RCCs were grateful that the infrastructure for this telementoring option was established as this allowed sites to continue with virtual platforms throughout the pandemic to meet timely needs.

- Pacific ECHO website: https://pacificscd.org

The Pacific Collaborative also implemented the following topic specific ECHOs:

- CBO ECHO
  - This CBO covered topics such as teaching other CBOs how to apply for funds
- Hemophilia (modified ECHO model)
  - Held monthly
- Insurance issues and practice guidelines specific to the SCD population
  - Hosted by Colorado
- Physical Therapy

Domain 3: Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

Improving Access to Quality Care

- Pacific sites have employed a number of strategies to improve access to quality care. For example, some sites have hired dedicated social workers, SCD health advocates, child life specialists, pediatric neuropsychologists, and child psychologists to address existing and emerging needs.
- Arizona increased support for coordination of care. This site conducts pre-visit planning with a dedicated nurse coordinator and a medical assistant who plans the SCD visits for the upcoming week so that visits with the patient are most productive.
- Washington has embedded a pediatric psychologist early in patients' care to normalize and reduce stigma of mental health support, specifically blocking psychologist appointment time.

Transition Planning

- In partnership with the Pacific RCC, the Sickle Cell Disease Foundation has created a nine-month transition program that works to ensure that adolescents are prepared for care transition. Through interactive program activities, the CBO/clinic partnership teaches participating adolescents how to manage the essential areas of SCD. At the end, the site holds a mock clinic with the medical team to which the new adult living with SCD will transfer. The adolescents are asked to rate the providers that they encounter, and the providers rate the encounters from their medical perspective. Based on this mock clinic, a shared decision is made about whether a young adult is ready to transition.
- The Colorado site has a strong transition program managed by social workers. The program includes implementing a youth advisory board and community interactions with the physicians to whom young adults living with SCD are referred.

- The Nevada site is in the process of launching *It's T.I.M.E. (Transitioning Into a more Mature Era)*. This program is designed for 18- to 25-year-olds who live with SCD and their immediate family members. Topics focus on life issues, such as finances, career options, family affairs, and education plans.

Partnership with Community-Based Organizations

- The Arizona clinical site forged a relationship with their local CBO, meeting for the first time at a Pacific RCC regional meeting. After meeting, they partnered to hold a full-day community event. The CBO took the planning lead, with the clinic providing educational support. This event has helped connect the clinic with the community, which had never been done. This CBO/clinic relationship remains strong and a link to bring together patients, families, and caregivers.

- The Pacific region has forged a model clinic/CBO relationship showing what true partnership can look like in SCD care. The RCC worked with the Sickle Cell Disease Foundation, the first and oldest nonprofit, social service, SCD organization in the U.S. Located in Southern California, the SCDF was started by four physicians and grounded the CBO in both the community and the clinic from the start. The SCDF is part of all the regional decision-making and the RCC values SCDF's input as a full partner. This has allowed the region to make great strides in maintaining connection with the community of people living with SCD. The Sickle Cell Disease Foundation conducted the following select activities:

    - Continued two long-time camp programs: one for teens called Camp Gibbous, and one for the younger children called Camp Crescent Moon. The camps provide opportunities to the children and teens who attend and to people living with SCD who work as camp counselors. Sixty percent of the camp counselors, aged from 21 to 40, are people living with SCD. It is an important experience for kids to see older people who live full lives with SCD. The teen camp is for adolescents aged 15-18 years and serves about 40 kids annually. The primary purpose of the camp is to help kids grow and understand that they can live with SCD. Participants learn how to advocate for themselves. Graduates of the camp include physicians, attorneys, and other highly-educated and trained people living with SCD. Many of them say that it was the camp that pushed them to say "I can," not "I can't." The camp continued during COVID-19 using a virtual platform to keep kids living with SCD and their families engaged and together.

    - Created a group specifically for people 21-30 years of age. This is often a "forgotten" group of adults, as most have transitioned out of pediatric care and are working, in college, or potentially starting a family. Since they started this group, they average between 20- 30 people on their Zoom calls.

MIDWEST REGIONAL COLLABORATIVE

Description of RCC Activities

Domain 1: Increase the number of providers treating individuals with sickle cell disease using the National Heart, Lung, and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report

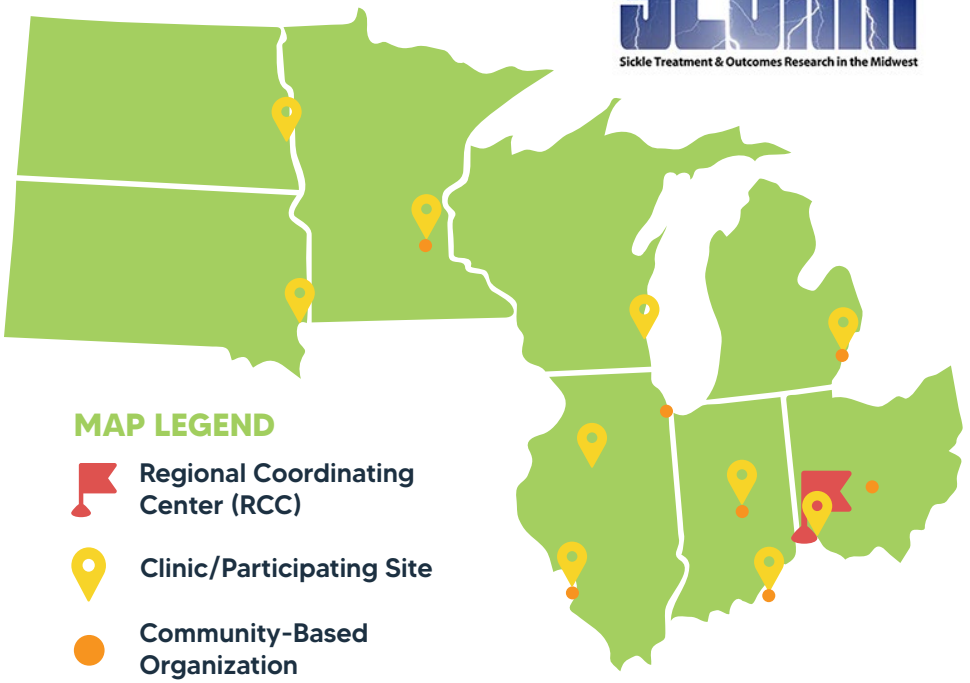
- Several Midwest sites worked on ways to improve rates of immunization, including improving team preparation for clinics; reviewing guidelines with staff; assigning a coordinator to track immunizations; hiring a pharmacist; inserting a maintenance tab in the EHR; implementing quick order sets (an organized list where the clinician can quickly mark off which immunizations are needed); and making past due immunizations visible to patients in their MyChart, a patient-facing medical portal.
- Several Program sites are participating in a multicenter HU study led by Cincinnati Children's Hospital Medical Center (CCHMC), the lead organization of the Midwest RCC. The Hydroxyurea Optimization through Precision Study (HOPS) is examining how best to optimize HU use (Meier et al., 2020). HOPS will answer important questions about the clinical feasibility, benefits, and safety of **pharmacokinetics-based guided dosing** of HU for children with SCD. Lessons from this study have the potential to change the treatment paradigm from a standard weight-based approach to one that safely and effectively optimizes the laboratory and clinical response in an individual person living with SCD.

Illinois

- Maintains a manually entered spreadsheet document that is updated quarterly and then rechecked at the beginning of each month because of EHR system limitations. This site has worked on clinical improvements for this health area since 2013. They are able to report on successes. At baseline, they had about a 50 percent completion rate. They now consistently sustain over an 80 percent, nearing 100 percent many months, up-to-date rate.
- Is implementing an HU quality improvement project to increase rates.

About the Midwest RCC

STORM
Sickle Treatment & Outcomes Research in the Midwest



Lead Organization

Cincinnati Children's Hospital Medical Center

Principal Investigator

- Lisa Shook, DHPE, MCHES

Data Manager

- Christina Bennett Farrell, MPH, CPM

Participating Clinics

- IL – Children's Hospital of Illinois-Peoria
- IN – Indiana Hemophilia & Thrombosis Center
- MI – Sickle Cell Disease Association of Michigan
- MN – Children's Minnesota

- Is expanding their Quality improvement for Urea Adherence in Kids with Sickle Cell Disease Study (QUAKS) to improve HU adherence among patients with SCD. The results from this 2018 QI project showed improved HU adherence and fostered health education/counseling, increased patient and family satisfaction, and enhanced service utilization. This site plans to continue to individualize care to increase adherence rates and sustain improvements.

Ohio

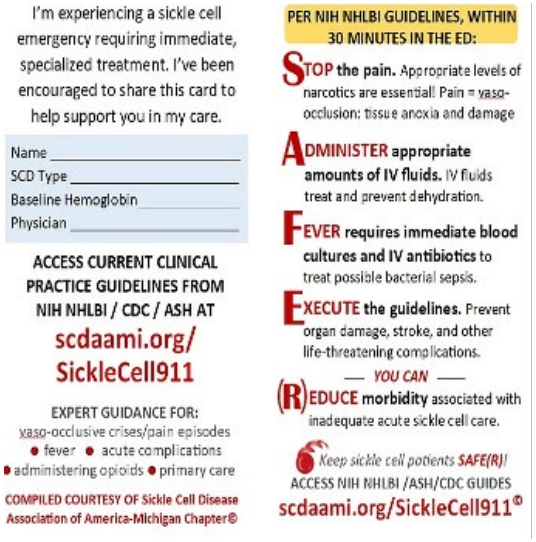
- Led an SCD Caregiver Immunization Survey across three regional sites to look at barriers to immunization completion.

- Is using QI data to improve HU adherence, reduce ED visits for pain, and prevent Acute Chest Syndrome.

Michigan

- Strengthened their relationship with a national association of emergency department doctors as these providers often see patients during moments of crisis and may not know the patient. Additionally, to help patients, this site developed SAFE(R) cards (Fig. 1) for people living with SCD to give to an emergency department doctor upon arrival. These cards are designed to provide a physician with information in the moment about the standard of care for taking care of people living with SCD when they arrive in the emergency room.

Fig. 1. SAFE(R) Card for People Living with SCD to Provide to ER Physician



Wisconsin

- Analyzes data quarterly, to identify patients due for a TCD. The report is provided to the nurses for feedback and scheduling. This team has also added a Child Life Specialist to assist in screenings of younger patients.

- Is conducting QI work with their electronic health record system to collect information for an SCD registry that will track quality indicators.

- ND – Sanford Health (Fargo)
- OH – Cincinnati Children's Hospital Med Center
- SD – Sanford Health (Sioux Falls)
- WI - Children's Hospital of Wisconsin

Participating Community-Based Organizations

- Sickle Cell Disease Association of American – Michigan Chapter
- Sickle Cell Disease Association of America – Illinois Chapter
- Martin Center (Indianapolis, IN)
- Sickle Cell Foundation of Minnesota (Minneapolis, MN)
- Ohio Sickle Cell and Health Association (Columbus, OH)

IMMUNIZATION EFFORTS

One hematologist with a long track record of strong immunization rates recounted the intense effort of the team under her direction. While her state has a statewide immunization database, physicians' offices are not required to participate, rendering that source unreliable. To create a reliable dataset, her data coordinator created an extensive Excel file for all critical immunizations.

To get complete information, her staff individually calls other physicians' offices to obtain faxed immunization records, which are then entered into their Epic EHR system. But because their Epic system cannot easily return patient-level information, time is also spent dually entering the data into the Excel database. Only the Excel database has all the pertinent information in a readily accessible format. This process is kept up monthly by a dedicated staff person.

Although cumbersome, it is the only way to complete the tracking, monitoring, and data collection since many of the specialist's patients are not within the hematologist's healthcare system.

Domain 2: Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care

RCC ECHO Website

RCCs found the Project ECHO® approach to be effective and manageable for knowledge-sharing. They report high use and impact of Project ECHO® and expressed desire for continued support to expand this capacity.

Using this model increased RCCs' ability to offer SCD-specific regional and national sessions. RCCs were responsible for planning, organizing, and facilitating ECHO sessions tailored to their regional needs and convening national ECHOs as needed. Content covered SCD-focused information, from basic science to psychosocial needs. ECHOs often focused on region-specific content and cases to best support providers where they served patients. However, all RCC ECHOs were "open" and people from any geographic location within the country and world were able to attend. Some recorded the education sessions to ensure the greatest reach possible. RCCs noted that making ECHO open to all, live and recorded, was an important service for providers in their regions for whom they were not able to educate more directly due to geographic or resource limitations. RCCs were grateful that the infrastructure for this telementoring option was established as this allowed sites to continue with virtual platforms throughout the pandemic to meet timely needs.

- Midwest ECHO website: https://sickleecho.org

The Midwest also sponsored COVID-19 specific ECHOs:

- Topics included medical and psychosocial impact of COVID-19 on children and adults living with SCD
- Hosted as needed with at least one session per month
- Attendee participation increased 150 percent during COVID-19 sessions

Additional Provider Teaching Opportunities

The Ohio site supported the annual national Hemoglobinopathy Counselor Training Course.

The Minnesota site developed a "Sickle Smart" webinar series. This free series offered a virtual classroom designed to provide information on treating the whole person living with SCD. The Sickle Smart webinar series planners used online lectures, discussion, and sharing of current resources and educational topics to strengthen participants' knowledge base about sickle cell disease, sickle cell trait, and the experience of people who live with SCD. The educational sessions were open to the public and designed for people living with SCD, their caregivers, healthcare providers, SCD advocates, educators, and others who want to learn about SCD. Importantly, it offered practical SCD resources for physicians, physician assistants, nurses, advanced practice providers, and other healthcare professionals. Recorded sessions are archived and can be accessed online.

Topics included:

- Complications and other issues related to SCD
- Current evidence-based practices to improve care of individuals with SCD
- New treatments recently available to individuals with SCD
- Challenges and improvement efforts related to transitioning individuals from pediatric to adult care
- Discussion of the difference between equality and equity and their impact on Health Equity and Social Justice
- Review of the MN Dept of Health's Newborn Screening and Long-Term Follow-up Programs and how they impact the work of improving outcomes for individuals living with SCD in Minnesota
- Resources available to individuals living with SCD

Domain 3: Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

Improving Access to Quality Care

Illinois

- Instituted the use of a tracking grid that includes all the items, including QI project issues, that need to be reviewed in a patient's pre-meeting. Every patient is discussed extensively: all medical issues (immunizations, TCD, transition, etc.), psychosocial needs, school-related issues, and preparation items for the visit with the patient and family. The team reaches out to the entities that have information about the patient (such as school-based health care clinics) so that care is covered comprehensively.
- Employed the use of regular calls from a nurse to engage with patients regarding questions about HU. These calls have built a trusting relationship over time. Now parents bring up other questions and concerns, including school, behavioral, and sleep issues.
- Grown their relationship with other hospital departments, such as their radiology division. The strength of this partnership has helped when a patient misses their TCD appointment. When the patient returns, the SCD team can alert radiology, who will complete the TCD. This has been instrumental in helping maintain strong usage of TCD and ensure comprehensive care.
- Conducted a missed appointment project that includes qualitatively interviewing patients and their families about reasons for missed care with the goal of improving appointment keeping.

Minnesota

- Has been a leader in addressing institutional racism. In 2020, their commitment was needed more than ever. In the wake of the murder of George Floyd, twelve blocks from their institution, community outrage brought civil unrest. Patients were frightened to come to the site. However, through their year's long commitment, this site had established the trust of the people they serve. This site regularly conducts presentations and corresponding discussions so providers and staff have a place to discuss race issues that impact their patients and the greater community in which they reside.

Transition Planning

The Midwest RCC has addressed and bolstered transition care in several ways. Some clinics have:

- Hired transition liaisons
- Increased the use of multidisciplinary teams that include a nurse coordinator and social worker
- Begun a local consortium to engage several adult providers and clinics to develop local guidelines and a patient-facing checklist to improve transition and evidence-based care for adults living with SCD
- Established procedures with adult medicine that ensures the transition of individuals aged 21 years and older to an adult SCD provider is completed
- Created automated reminders for appointments and built EHR entry fields to make sure care is offered in a timely way and to track whether or not care is completed.

ADDRESSING PSYCHOSOCIAL NEEDS

Comprehensive programs, such as the CASCADE program in Indiana, highlight RCC commitment to achieving high rates of national standard care. The Indiana site, The Indiana Hemophilia and Thrombosis Center (IHTC), has worked with their State Department of Health to start Community Access for Sickle Cell ADult CarE (CASCADE), aimed at addressing the needs of Indiana adults aged 21+ living with SCD related to mental health, vocational rehabilitation, and pain management.

This project has a three-fold purpose:

- To increase access to evidence-based care for adults living with SCD
- To collect and utilize data to target services to those who need them most
- To expand educational outreach about SCD and sickle cell trait

- Established satellite clinics to address transportation issues. Specifically, Illinois has leveraged Program funds to enhance their rural SCD outreach clinics while Indiana has been expanding their northwestern outreach clinics.

In 2018, Minnesota worked with their department of health to develop a transition toolkit. This pediatric site works with University of Minnesota's adult program to ensure patients are ready for care transition. The toolkit prescribes a timeline for activities: what age to start conversations; setting parent expectations; teaching children to be able to make their own appointments by a certain age; having a set plan for picking up medications; teaching the young person how insurance works, etc. Working through this toolkit has helped the site be more efficient with determining role division between the nurse case manager, the SCD health advocate, and the social worker, helping make sure all topics are covered and eliminating duplication.

PASSPORTS TO HEALTH

Thinking about how difficult it is to get doctors to take care of SCD patients, the Michigan CBO site created a patient empowerment toolkit, or what they call a "passport to health." This toolkit helps people living with SCD take better care of themselves by engaging them in their own healthcare.

The "passport" is used by community health workers with patients to make sure they know what type of SCD they have, their usual hemoglobin level, and dosages of medications (and how to spell them) so that when patients are sent to new doctors, they feel empowered and are ready to participate in client-centered care. As part of that toolkit, the site created a seven-minute white board video. This format was chosen to keep patients engaged. Many people living with SCD have cognitive and frontal lobe issues that can make it difficult to retain and take in large amounts of written information.

Partnership with Community-Based Organizations
Minnesota used some of their resources to hire an SCD family health advocate who is a critical component of community support for patients and families and a partnership brought about by their involvement with the STORM RCC. This site's missed appointment rate has significantly decreased.

The Michigan CBO:

- Invites mothers who have babies diagnosed with SCD to a series of educational sessions that allow participants to interact and support each other and leave with a system of support in addition to important information about SCD. These meetings allow families to talk to each other, allowing caregivers to collectively process the call they received telling them their baby was born with SCD. Parents exchange phone numbers, and they receive a briefcase to keep all their child's information. By the end, they know they are not alone and have a built-in support system if they want it.

- Has hired four patient advocates who can offer support for both people living with SCD and with SCT. Patient advocates and community health workers strategically placed in Black communities along with satellite offices cover the entire state. This CBO also offers group and individual counseling, attends health fairs to offer SCD testing, and provides education on SCD, SCT, and related issues.

References

Meier, E. R., Creary, S. E., Heeney, M. M., Dong, M., Appiah-Kubi, A. O., Nelson, S. C., . . . McGann, P. T. (2020). Hydroxyurea Optimization through Precision Study (HOPS): Study protocol for a randomized, multicenter trial in children with sickle cell anemia. *Trials*, 21(1), 983. https://doi.org/10.1186/s13063-020-04912-z

HEARTLAND/SOUTHWEST REGIONAL COLLABORATIVE

Description of RCC Activities

Domain 1: Increase the number of providers treating individuals with sickle cell disease using the National Heart, Lung, and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report

- Iowa, Arkansas, and Missouri developed innovative, team-based approaches to improve immunization rates among their patients. Specific activities included:
  - Organizing comprehensive teams with providers from varying disciplines
  - Providing extensive education on the need for immunizations among people living with SCD
  - Reviewing timelines for distribution
  - Planning pre-clinic huddles
  - Developing clinic prep sheets
- Worked with their IT departments to develop immunization order sets for patients with SCD in their EHR systems
- Developed algorithms for common issues arising in SCD patients (i.e., timely treatment of vaso-occlusive crisis, use of intranasal fentanyl EHR order sets), post-discharge communication, and follow-up

Arkansas

- Overhauled their entire immunizations process to improve rates. This new process included:
  - Working with their health department, which has a well-established immunization database that pulls directly into their medical record system. Once this connection is established, data reflected vaccination status no matter where they are given
  - Creating a new EHR order set that established an internal system to review immunization needs prior to each appointment
  - Changing clinic flow to allow doctors to order vaccines prior to clinic visits so vaccines could be pre-stocked in their medication

About the Heartland/Southwest RCC

Lead Organization
Washington University School of Medicine, St. Louis

Principal Investigator
- Allison King, MD, PhD, MPH

Data Manager
- Taniya Varughese, MSOT, OTR/L

Participating Clinics
- AR - University of Arkansas for Medical Sciences, Arkansas Children's Hospital and Research Institute
- IA - University of Iowa Stead Family Children's Hospital
- KS - University of Kansas Medical Center
- LA - Louisiana State University Pediatrics, New Orleans Children's Hospital

dispensing system. The nursing teams were then able to release orders as soon as the child arrived to clinic. With this process, blood is drawn and vaccines are given before patients are seen by the physician.

Iowa

- Conducted an innovative project on pain assessment with their pediatric population. They distributed the Youth Acute Pain Functional Ability Questionnaire (YAPFAQ), a self-report measure of physical function in youth experiencing acute pain. Results from this assessment help providers better understand how pain is functionally limiting a patient (e.g., making it difficult to complete activities of daily living, falling asleep, listen to providers). Early findings show:
    - 93 percent of pediatric patients (n=14) are knowledgeable about YAPFAQ and its purpose
    - 70 percent of nurses noted that YAPFAQ made a difference in the pain management they provided to pediatric patients

Missouri (St. Louis Children's Hospital)

- Developed a QI project to improve pain management for inpatients with SCD to reduce time to treatment and increase satisfaction with pain management. For this project they:
    - Consulted pain management services
    - Increased the use of non-opioid adjuvant pain medications

Texas

- Established a collaborative process with their radiology team so that TCDs are always scheduled with a general SCD appointment. Patients receive their TCD first so that the result is ready by the time they are seen for their general appointment.

- Overhauled its data system to be able to track HU prescription rates.

Domain 2: Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care

State Plans

For the Program, RCCs developed a Regional Sickle Cell Action Plan and state-specific Sickle Cell Action Plans for funded states. The plans included 1) resources in each region and state to improve SCD care for all people with

- MO - Washington University School of Medicine, Barnes Jewish Hospital, St. Louis. Children's Hospital, Truman Medical Center, Missouri University Health Care
- NE - University of Nebraska Medical Center, Children's Hospital and Medical Center
- OK - University of Oklahoma Health Sciences Center
- TX - Baylor College of Medicine, Texas Children's Hospital

Participating Community-Based Organizations

- Sickle Cell Association (St. Louis)
- St. Louis Integrated Health Network
- Sickle Cell Association of South Louisiana and Baton Rouge Sickle Cell Anemia Foundation
- Supporters of Families with Sickle Cell Disease (Oklahoma)
- Sickle Cell Association of Texas, Marc Thomas Foundation

SCD in the region and 2) a description of the overall infrastructure that would address the goals and requirements listed in the FOA.

The state action plans described:

- How each state intended to develop a network of providers using evidence-based SCD care in the state
- How telemedicine/telehealth strategies and other provider support would be utilized
- How access to quality care would be supported
- How the state intended to increase the number of individuals with SCD being treated by providers using evidence-based SCD care

These plans were used to help formulate a cohesive approach to facilitate knowledge-sharing. Discussions spurred by these state plans brought forth a regional vision and strong partnerships with the RCCs that continues today. Information included may be helpful to states interested in similar work.

Heartland/Southwest's Plan

RCC ECHO Website
RCCs found the Project ECHO® approach to be effective and manageable for knowledge sharing. They report high use and impact of Project ECHO® and expressed desire for continued support to expand this capacity.

Using this model increased their ability to offer SCD-specific regional and national sessions. RCCs were responsible for planning, organizing, and facilitating ECHO sessions tailored to their regional needs and convening national ECHOs as needed. Content covered SCD-focused information, from basic science to psychosocial needs. ECHOs often focused on region-specific content and cases to best support providers where they served patients. However, all RCC ECHOs were "open" and people from any geographic location within the country and world were able to attend. Some recorded the education sessions to ensure the greatest reach possible. RCCs noted that making ECHO open to all, live and recorded, was an important service for providers in their regions for whom they were not able to educate more directly due to geographic or resource limitations. RCCs were grateful that the infrastructure for this telementoring option was established as this allowed sites to continue with virtual platforms throughout the pandemic.

- Heartland/Southwest ECHO website: https://sicklecell.wustl.edu/scd-teleecho-clinic-164

Additional Provider Teaching Opportunities
- Several Heartland/Southwest sites partnered with the American Society of Hematology and their Clinical Trials Network, including participation with SCD-centered workshops.
- Heartland/Southwest PI provided bi-monthly consultation to providers on patients with SCD through Centene, a healthcare network that delivers services across all 50 states.

Domain 3: Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

Improving Access to Quality Care
- One Missouri site piloted an SCD-focused, shared medical appointment process, modeled after a diabetes self-management program, to increase patient understanding of disease self-management and facilitate engagement and trust. Patients had access to a psychologist, pharmacologist, and a nurse practitioner during the appointment.
- Other sites in the Heartland/Southwest started to bundle appointments together. Although their clinic appointments are longer, patients can now see the hematologist, pulmonologist, social worker, as well as get their TCD and necessary immunizations all in one visit. Patients and their families have been very pleased with the efficiency of services and decreased need for multiple clinic visits.

Transition Planning
Several sites worked to improve care transition planning. For example, select regional sites offered care transitioning patients tours of the adult clinic and adult hospital services, and meet and greets with the adult providers prior to formal care transition. Pediatric sites have been more hands-on in the care transition process and continue to offer care coordination to help adolescents schedule their first appointments in the adult clinic and provide check-ins to make sure patients successfully made it to their adult appointments.

- The Heartland/Southwest RCC PI held a weekly young adult transition clinic to help ease the transition process for young adults aged 18 to 26 years. Because the young adult clinic is smaller, the team can offer personalized care, especially spending time to address self-management skills and psychosocial stressors that may impact their success in managing their disease independently. Since 2017, over 50 individuals have passed through the young adult clinic, with many successfully transitioning to the adult clinic upon matriculation.
- The Heartland/Southwest RCC hosts a regional website and routinely updates it with a list of adult providers throughout the region who are accepting new patients.
- A Heartland/Southwest RCC occupational therapy student developed a life skills and transition self-management program founded on implementation science principles. The program has discrete modules addressing pertinent areas of need (disease education and awareness, self-efficacy and communication skills, and adult self-management skills) for adolescents and young adults living with SCD. This program is currently being piloted solely through telehealth. Interim analysis and next steps include:
    - The 5 participants who have completed the program report significant increases in SCD-related self-efficacy and improvement in SCD knowledge and transition readiness skills post intervention.
    - This program will continue to recruit more participants and Washington University published the modules and protocol for national dissemination and plans to adopt this work into standard of care.
- Oklahoma created an EHR template for improving documentation for care transition planning. They also designated a nurse practitioner and social worker to lead a "Sooner Success" Health Care Transition Committee to pilot a study to improve rates of pediatric patients with written care transition plans and documented care transition skills.

Partnership with Community-Based Organizations

- The Oklahoma CBO successfully forged partnerships with local and state agencies, including the Oklahoma Health Care Authority Initiative and Oklahoma Sickle Cell Collective Impact Team, to provide community awareness, advocacy, outreach, and comprehensive community-based care for people living with SCD throughout the state.
    - As a funded grantee of the SCDAA HRSA Newborn Screening Program, the CBO was able to train dedicated community health workers to engage unaffiliated patients and connect them to clinical care.
- The Texas CBO worked closely with the RCC state lead to provide more than 750 clients with:
    - Certified case management referrals to assist with social security and disability applications and renewals, housing, and transportation
    - Counseling, transition services, care coordination, medical home placement, scholarship programs, and other financial assistance
    - Organization of several SCD-focused camps (Camp Cell-A-Bration, ESCAPE, and Camp Next Level)
    - Support group meetings and free sickle cell trait testing
    - Organization of community awareness and advocacy events, including conferences and walks.
- The St. Louis CBO regularly hosted community education and awareness events, including walks, provider panels, and discussions on the importance of participation in research. The St. Louis CBO also:
    - Offered monthly support groups and real-time advocacy services to patients who were in the ED or inpatient at local hospitals
    - Mentored smaller CBOs throughout the region to develop effective programming for their patients
    - Collaborated with medical providers at local institutions to champion the development of three task forces to improve access to quality care for people living with SCD, focusing on:
        - Improving transition readiness for adolescent patients
        - Improving pain management in the ED
        - Working with legislators to inform policy changes for SCD

SOUTHEAST REGIONAL COLLABORATIVE

Description of RCC Activities

Domain 1: Increase the number of providers treating individuals with sickle cell disease using the National Heart, Lung, and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report

Alabama

- A Southeast RCC Co-PI is leading a project which is evaluating novel strategies to improve stroke screening for children living with Sickle Cell Disease. Dissemination and Implementation of Stroke Prevention Looking at the Care Environment (DISPLACE) Study (Kanter, NCT04173026). Data are anticipated to be released June 2022.
    - Several SE RCC sites participate with this study, thus implementing strategies with their current SCD population. See here for additional information about this study and its findings. (https://www.uab.edu/medicine/sicklecell/research/displace-trial). In order to achieve a "stroke-free generation," the goals of DISPLACE are to:
        - Assess the gap between current and recommended evidence-based practice for stroke prevention in children with SCD
        - Evaluate the effectiveness of a novel, multi-level dissemination and implementation strategy that engages providers, patients, and healthcare systems
        - Use engagement to enhance implementation of annual TCD screening and chronic red cell transfusion initiation at participating institutions for people living with SCD

Georgia

- One site changed their scheduling practices to try and improve TCD completion. This site began identifying those who needed a TCD each week and sent the list directly to the appointment scheduler. This site complemented this procedure change by following up with all missed appointments. With these changes, the site saw an increase in TCD completion rate from 42 percent to their current 74 percent.

About the Southeast RCC

THE EMBRACE NETWORK

Lead Organization

Levine Cancer Institute, Atrium Health

Principal Investigators

- **Julie Kanter**, MD, Director, Adult Sickle Cell Program | Co-Director, Lifespan Comprehensive Sickle Cell Research Center Hematology/ Oncology UAB Medicine | University of Alabama at Birmingham
  Co-Principal Investigator
- **Ify Osunkwo**, MD, MPH, Director, Adult Sickle Cell Disease Enterprise, Atrium Health
  Co-Principal Investigator
- **John Strouse**, MD, PHD, Director of the Adult Sickle Cell Program | Associate Professor of Medicine and Pediatrics, Duke University School of Medicine
  Co-Principal Investigator

Kentucky

- Kentucky began standard HU use in 2012 with children 9-12 months. This site compared the ED visit rate from 2012 to 2015. Between those time points there was a greater than 50 percent reduction in ED visits with that intervention. This site continues to follow this practice and has maintained these gains to this day.

South Carolina

- South Carolina is conducting QI work that looks at disease-modifying therapy, tracking TCD, and the benefit of pre-appointment checklists.

Domain 2: Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care

RCC ECHO Website

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Data Manager

- Shirley H. Miller

Participating Clinics

- AL - University of Alabama at Birmingham
- FL - University of Miami
- GA - Augusta University, Emory University/CHOA
- KY - University of Louisville
- MS - University of Mississippi
- NC - Duke University, Atrium Health
- SC - Prisma Health
- TN - **no current state lead

Participating Community-Based Organizations

- Piedmont Health Services and Sickle Cell Agency (North Carolina)
- Bridges Pointe, Inc. Sickle Cell Foundation (North Carolina)
- The Sickle Cell Foundation (Central Alabama)
- SCDAA – Miami-Dade County Chapter, Inc. (Florida)
- SCDAA – St. Petersburg Chapter, Inc. (Florida)
- Sickle Cell Foundation, Inc. – Tallahassee (Florida)
- Sickle Cell Foundation of Kentuckiana (Kentucky)
- Sickle Cell Foundation of Georgia, Inc.
- Huisman Sickle Cell Foundation of Augusta, Georgia
- James R. Clark Memorial Sickle Cell Foundation (Columnia, South Carolina)

- Southeast's ECHO website is under development
- Southeast-specific ECHO clinics include:
    - Curative therapies (national audience)
        - Hosted by Emory twice a month
        - Aimed at hematology and transplant clinicians
    - Psychosocial
        - Hosted by Atrium Health

Additional Provider Teaching Opportunities

- The Georgia (Augusta) site conducted a patient satisfaction survey looking at satisfaction with subspecialty medical care delivered via telemedicine clinics (Stone et al., 2019). Results included:
    - Telemedicine experience was demonstrated positive, with patient's rating their experience at an average score of 3.79 out of a total of 4 points, for the 10 questions
    - A majority of families (42.5%) preferred telemedicine consultation to onsite physician visits; 91.3% would use telemedicine again; 89% would recommend telemedicine to others
    - Conclusions: Patients reported positive experiences with telehealth technology, staff, and hematologists. Telemedicine is a feasible approach to enhance access to hematology medical care for SCD
- Select site leads (from both the Northeast and Southeast RCCs) have been chosen to co-lead the American Society of Hematology-sponsored SCD center training workshops which occurred in 2019 and will be held again in 2021.
- Some Southeast sites implemented a system in an adult program that requires that every SCD patient admitted gets assigned to the hospitalist's service. With this revised system, **hospitalists** uniformly used one standardized inpatient order set for adults and a standardized order set in the ED. This provided consistency and helped providers feel comfortable treating patients. The lead PI of this site noted that with this change, the hospitalists only need to reach out to the SCD specialist for specific questions regarding patient care.
- South Carolina developed a transition and mentor programs

Domain 3: Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

Improving Access to Quality Care

- The Georgia sites established phone-based telemedicine over 3 years ago and have provided transportation assistance to reduce barriers to care.
    - These sites relied on strong partnership to make this happen. State leadership showed tangible support by supplying equipment and the staffing needed to make telemedicine visits possible early on. These groups also worked together to ensure that billing for these types of visits was reimbursable.
    - Nursing teams assist with scheduling, HIPAA compliance, intake, and tasks that were initiated with the start of telemedicine.
- In Kentucky, the addition of a care coordinator freed time for clinicians and ensured patients were linked to the psychologist and the social worker who could better support psychosocial needs.
- The South Carolina site is a large clinic that serves pediatric and adult patients. It established a comprehensive SCD specialty clinic which provides:
    - Red blood cell exchange on-site services for patients (pediatrics and adults) who otherwise may have to travel to multiple locations for care
    - Full-service TCD capacity, which was necessary for strong completion rates
    - Pain management services in the hospital and as outpatient services that address the bias issues that patients often face when seeking pain management in EDs or pain clinics that do not have SCD experience
    - Access to specialty medical providers, social workers, nursing psychiatry, research staff, sleep medicine, and child life specialists

PATIENT ENGAGEMENT DURING COVID-19

Southeast Atrium: Created Zoom in N2U engagement and education sessions (PEEPs). This well-attended series covered an array of COVID-19 related topics.

Table 1. Patient Engagement Education Sessions During COVID-19

PATIENT ENGAGEMENT DURING COVID-19			
SESSION	EDUCATION SESSIONS	DATE	ATTENDEES
1	**SCD in the Era of Coronavirus (COVID-19) What You Need to Know! Pt. 1**	3/18/2020	97
2	**SCD in the Era of Coronavirus (COVID-19) What You Need to Know! Pt. 2**	4/1/2020	75
3	**SCD in the Era of Coronavirus (COVID-19) What You Need to Know! Pt. 3**	4/8/2020	80
4	**(COVID-19) Q&A Session with the Sickle Cell Team & Dr. Ify**	4/15/2020	49
5	**Q&A Session with the Sickle Cell Team & Dr. Ify**	4/22/2020	94
6	**Let's Talk About Stress, Anxiety, COVID-19, & Coping Skills**	4/29/2020	66
7	**PART 1: NC State Re-opening... What Does That Mean for Someone Living with SCD?**	5/13/2020	57
8	**PART 2: NC State Re-opening... What Does That Mean for Someone Living with SCD?**	5/20/2020	65
9	**What Does the New Norm Look Like for Someone Living with SCD**	5/27/2020	35
10	**Fear • Anger • SCD #BlackLivesMatter**	6/10/2020	87
11	**Q&A SCD & COVID-19**	7/1/2020	
12	**What is Your Sickle Cell Action Plan During This Pandemic?**	7/29/2020	75

- Florida instituted a missed appointment protocol thereby reducing these occurrences
    - The site uses reminder calls the evening before and morning of TCD appointments, which has improved TCD completion rates and invested in patient-centered care and relationship building

Transition Planning

- Most states in the Southeast RCC have started recruiting for the ST3P-UP Transition Study. One of the RCC leads, Dr. Ify Osunkwo, a grant called ST3P-UP to enhance transition and engage community partners. This study is comparing the effectiveness of adding virtual peer mentoring to a transition program in improving acute care reliance, quality of life, and satisfaction with the care transition process in young adults living with SCD. The care transition program is based on the six core elements of Got Transition®. (https://www.pcori.org/topics/transitional-care/lessons-learned/engaging-community-partners-research-studies). The results are expected in May 2023.

- One of the Georgia sites is committed to addressing transition planning as a primary effort. They will share what they learn with the other GA clinics to ensure spread of best practices within this program.

- South Carolina has established care transition planning as a primary QI project. An initial review revealed that few care transition plans were documented. Since that time, the site has initiated a care transition readiness program that includes:
    - Beginning the process at age 12; confirming that medical consent policy has switched over to the young adult by age 16; and a final goal to a full adult care model by age 18
    - Tracking completed educational pieces are in clinics EHR
    - Now, chart audits show that this site has 90 percent or more of patients with a transition plan in place and who had a readiness survey completed within the past six months (what the site uses as another success measure).

Partnership with Community-Based Organizations

- One SE CBO schedules regular monthly meetings for patients, where they can share concerns and needs. And the CBO invests in the community through their sponsorship of a fundraiser for scholarships for high school seniors who have SCD.

- Kentucky did not have an established relationship with the local CBO at the beginning of this most recent program period. With support from the Program, this site made improving communication and engagement a priority. The site hired a social worker with time dedicated to the Program in order to grow this relationship. Today, the clinic and the CBO view themselves as a team. With more than 70 referrals from the clinic to the CBO in the past two years, the clinic has seen the impact that the connections make on a patient's life. Because of the strong partnership, patients now communicate directly with the CBO to receive assistance or support. The evolution of this relationship has been important in educating and advocating within the clinic administration about the importance of easier access to the clinic and to patients.

References

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Stone, R., Chung, Y., Stone, K., Ameri, A., & Pace, B. S. (2019). Telemedicine expands hydroxyurea monitoring for children living with sickle cell disease in rural south Georgia. *Biomedical Journal of Scientific & Technical Research*, 22(1), 16380-16385.

NORTHEAST REGIONAL COLLABORATIVE

Description of RCC Activities

Domain 1: Increase the number of providers treating individuals with sickle cell disease using the National Heart, Lung, and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report

- One NY site initiated an ED QI project with the goal to have patients who go to an ED affiliated with the hospital network to have an individualized pain protocol available in the EHR system or a standardized pain protocol when no individualized protocol is available. The site is trying to reduce the reliance on ED teams to have to make decisions about patients they may not know. An extended goal is to reach additional neighborhood hospitals so that ED staff can review and use existing pain protocols. The current protocol calls for connecting patients with a community health worker to facilitate linkage to the local sickle cell center after the ED visit as well. This process has been completed at one hospital.

Domain 2: Use telementoring, telemedicine, and other provider support strategies to increase the number of providers administering evidence-based SCD care

RCC ECHO Website

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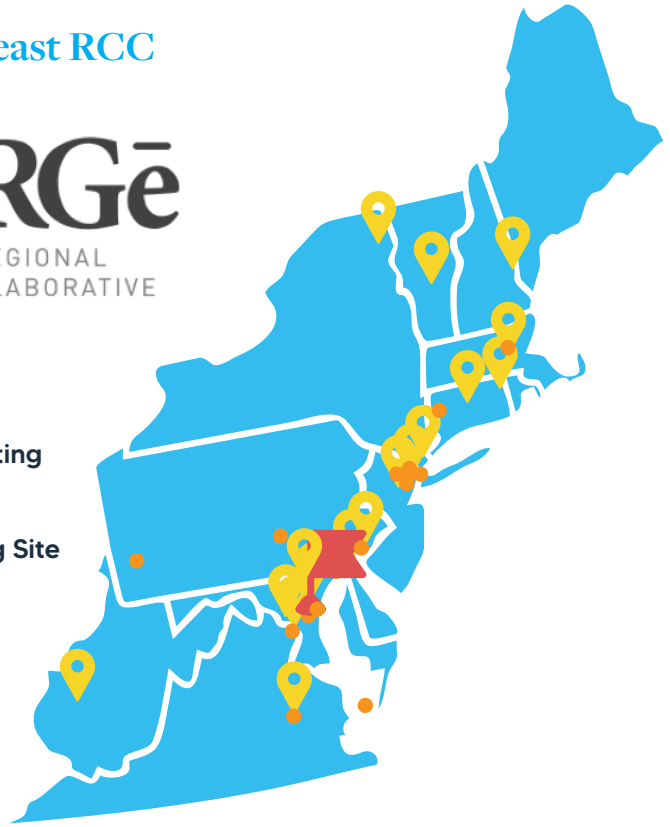
Using this model increased their ability to offer SCD-specific regional and national sessions. RCCs were responsible for planning, organizing, and facilitating ECHO sessions tailored to their regional needs and convening national ECHOs as needed. Content covered SCD-focused information, from basic science to psychosocial needs. ECHOs often focused on region-specific content and cases to best support providers where they served patients. However, all RCC ECHOs were "open" and people from any geographic location within the country and world were able to attend. Some recorded the education sessions to ensure the greatest reach possible. RCCs noted that making ECHO open to all, live and recorded, was an important

About the Northeast RCC

SiNERGē
NORTHEAST REGIONAL SICKLE CELL COLLABORATIVE



Lead Organization

Johns Hopkins University

Principal Investigators

- Sophie Lanzkron, MD, MHS
  Co-Principal Investigator
- Rosalyn Stewart, MD, MS, MBA
  Co-Principal Investigator

Data Manager

- Bailey House, MPH

Participating Clinics

- CT – University of Connecticut
- DE – Tova Health
- District of Columbia – Howard University
- MA – Boston Medical Center

service for providers in their regions for whom they were not able to educate more directly due to geographic or resource limitations. RCCs were grateful that the infrastructure for this telementoring option was established as this allowed sites to continue with virtual platforms throughout the pandemic to meet timely needs.

- Northeast ECHO website: https://www.hopkinsmedicine.org/Medicine/sickle/

- The NE created five ECHO clinics:
    1. **Children's Sickle Cell Foundation:** Community-based Organizations in SCD community focused on mentorship and education
    2. **Jacobi Medical Center:** Quality Improvement; Projects related to ED, stigma, registry implementation, and transition; teaching attendees about the QI process
    3. **BMC/Rhode Island Hospital:** Pediatric Patient Care
    4. **Johns Hopkins University:** Adult and Pediatric Patient Care
    5. **Virginia Commonwealth University:** Adult Patient Care
- Through these clinics, the NE offered:
    - 276 sessions, resulting in 327 hours of education/mentoring. There were 162 separate educational sessions with 279 cases presented.
    - Logged more than 200 hours of continuing medical education (CME) and maintenance of certification credits — both huge incentives needed to encourage providers to participate. In total, 3,372 attended, with more than 559 participants.

Additional Provider Teaching Opportunities

- Launched a recurring educational series in collaboration with Globin Research Network for Data and Discovery (GRNDaD) called "GRNDaD Speaks! SCD Speaker Series." Local and international expert speakers presented basic science research in SCD.
    - In 2020 this series completed 21 sessions with 2029 attendees.
    - The plan is to continue this series indefinitely.
- Select site leads (from both the Northeast and Southeast RCCs) have been chosen to co-lead the American Society of Hematology-sponsored SCD center training workshops which occurred in 2019 and will be held again in 2021.

- MD – Johns Hopkins University
- ME – Maine Children's Cancer Program
- NH – Dartmouth-Hitchcock Medical Center
- NJ – Newark Beth Israel Medical Center
- NY – Jacobi Medical Center
- NY (2nd lead) – Columbia University Medical Center
- PA – Children's Hospital of Philadelphia
- Puerto Rico – Universidad de Puerto Rico
- RI – Rhode Island Hospital
- US Virgin Islands – Virgin Islands Oncology and Hematology
- VA – Virginia Commonwealth University
- VT – University of Vermont Children's Hospital
- WV – Charleston Area Medical Center

Participating Community-Based Organizations

- Citizens for Quality Sickle Cell Care* (Connecticut)
- Sickle Cell Association of Delaware
- William E. Proudford Sickle Cell Fund, Inc. (Delaware)
- Faces of Our Children (District of Columbia)
- Sickle Cell Association of the National Capital Area, Inc. (DC)
- Armstead-Barnhill Foundation for Sickle Cell Anemia (Maryland)
- Association for the Prevention of Sickle Cell Anemia Harford and Cecil Counties and the Eastern Shore* (Maryland)
- Christopher Gipson Sickle Cell Moyamoya Foundation (Maryland)
- Maryland Sickle Cell Disease Association*
- William E. Proudford Sickle Cell Fund, Inc. (Maryland)
- Greater Boston Sickle Cell Disease Association* (Massachusetts)
- Sickle Cell Association of New Jersey*
- Candice Sickle Cell Fund, Inc. (New York)
- Queens Sickle Cell Advocacy Network* (New York)
- Sickle Cell Awareness Foundation Corp International (New York)
- Sickle Cell/Thalassemia Patients Network* (New York)
- Children's Sickle Cell Foundation Inc.* (Pennsylvania)
- SCDAA – Philadelphia/Delaware Valley Chapter* (Pennsylvania)
- South Central Pennsylvania Sickle Cell Council* (Pennsylvania)
- Anemia Falciforme Sickle Cell Disease en Puerto Rico (Puerto Rico)
- Life and Family Foundation Richmond (Virginia)
- Sickle Cell Association Inc.* (Virginia)

**Chapters of the SCDAA*

Domain 3: Develop and implement strategies to improve access to quality care with emphasis on individual and family engagement/partnership, adolescent transitions to adult life, and care in a medical home

Improving Access to Quality Care

- To ensure each patient receives similar guideline care for both screening and follow-up, one rural site made the decision to centralize care of SCD patients to one hematologist/oncologist.

Transition Planning

- Select locations of the NY Health + Hospital System have been working to start transition care planning early. These locations are piloting a formal transition-of-care program using the *Got Transition*® program. Introducing the program to patients when they turn 12, the program includes:
    - Dedicating two of four annual visits to topics related to transition to adult care
    - Striving to integrate the early transition planning into the hospital system by adding it to young people's medical records

  The end goal is to have the entire health system commit to this program post pilot.

Partnership with Community-Based Organizations

- A lead CBO convened local CBO leaders monthly to collaborate and work collectively. The group discussed collective work and goals. This built capacity and leadership that is needed to sustain strong regional CBO work moving forward.

- Supported by the Program, one of the regional CBOs focused on capacity-building, collaboration, and creating space for CBO leadership. They offered a small grants program to engage and support CBOs.

- One of the CBOs started a specific CBO ECHO.

Appendix B: Sickle Cell Disease Overview & Background

REPORT TO CONGRESS
SEPTEMBER 2021

Section 1 | Incidence and Prevalence

Sickle Cell Disease

Approximately 100,000 Americans live with SCD. There is no precise estimate of SCD global incidence. Current estimates indicate that worldwide close to 300,000 people are born with SCD each year. Measuring a more precise prevalence of the disease would be helpful in ensuring all patients are receiving adequate care. Without surveillance data that track the true prevalence of SCD in the U.S. and territories, understanding where patients live and where they are receiving care — or if they even seek care — is unknown. Better surveillance is imperative to identify areas of concern and address gaps in care.

Sickle Cell Trait

While people with SCT do not have the symptoms or illness of SCD, they carry the sickle cell gene and, if their partner also carries the sickle cell gene, there is a chance their children may have sickle cell disease. Partnership with the local and state newborn screening programs is important to ensure that comprehensive counseling is conducted so that people can make informed decisions about family planning.

More than 1.1 million newborn babies were born with sickle cell trait (SCT) in the United States (Benson & Therrell Jr, 2010).

- This means one in nearly 70 babies have SCT (Kato et al., 2018)
- 1 in 13 Black or African-American babies is born with SCT.
- More than 100 million people live with SCT around the world.

ABOUT SCD TYPES

Hemoglobin SS disease is the most common type of sickle cell disease. It occurs when a person inherits copies of the hemoglobin S gene from both parents. This forms hemoglobin known as Hb SS. As the most severe form of SCD, individuals with this form also experience the worst symptoms at a higher rate (Healthline, 2019).

Severe Physical Complications

In addition to the complications described in the main report, listed here are additional severe complications.

- SCD also causes damage to kidneys, the liver, and other organs.
- One-third of male adolescents and young men with SCD suffer from painful, prolonged erections (priapism), which can result in scarring, deformation, and impotence (Idris et al., 2020).
- Girls and women with SCD may have delayed puberty, more intense pain before and during menstruation, difficulty getting pregnant, or complications during pregnancy (Andemariam & Browning, 2013; Ghafuri et al., 2017; Kuo & Caughey, 2016; Stimpson et al., 2016).
- More information about acute and chronic complications from SDC can be found in the *National, Heart, Lung and Blood Institute Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014.*

Minimizing physical symptoms and disease progression of SCD is important to patients; to achieve this, healthcare costs for patients with SCD are substantial. In 2016, it was estimated that the average person living with SCD accrued nearly $1 million in total lifetime healthcare costs, with annual costs of more than $30,000 for adults (Novartis, 2019). This is before adding two new essential medications, Crizanlizumab and Voxelotor, whose prices — although potentially covered by patients' insurance — range from $7,000-10,000 a month (BioPharma Dive, 2019; CVS Health, 2020). Addressing this issue is especially important given that SCD is a lifelong condition that impacts several health areas.

Section 2 | SCD Evidence-Based Care: Therapies, Disease Prevention, Screening, Pain Management

Consistent access to high-quality care profoundly affects outcomes for people living with SCD. For example, detection of affected individuals at birth and interventions, such as penicillin prophylaxis, vaccines, aggressive response to fever and administration of broad spectrum antibiotics (Brousseau et al., 2010; National Heart Lung and Blood Institute, 2002), blood transfusion protocols, introduction of medications to control condition symptoms, and Transcranial Doppler (TCD) screening (Section on Hematology/Oncology Committee on Genetics, 2002) have contributed to dramatic improvements in quality of life and life expectancy. However, some of these treatments require specialized clinical settings and not all patients have equal access to these types of care. Additionally, according to the Office of Minority Health, only approximately 1 in 4 people living with SCD receive the standard of care outlined in current National Lung Blood and Hemoglobin Institute *Evidence-Based Management of Sickle Cell Disease (SCD) Expert Panel Report* (Office of Minority Health, 2020), which provides standard primary guidelines for improving care.

Therapies

The pathophysiology of SCD is well understood and development of medication has occurred over the last several years. The collective goal of available medications is two-fold: first, to improve quality of life for patients by reducing the incidence of common SCD-related complications, such as pain crises and acute chest syndrome; and, second, to reduce the associated emergency department visits and hospitalizations, which will lower overall costs of care (Wang et al., 2013). Below are medications that have been developed.

Medication

Hydroxyurea (HU, aka Siklos, Addmedica, Droxia)

HU was approved by the U.S. Food and Drug Administration (FDA) for adults with SCD in 1998, but not for children until 2017. HU is a significant SCD therapy option. In SCD patients, it has been effective in reducing the frequency of pain crises and need for blood transfusions in children 2 years and older (National Heart Lung and Blood Institute, 2014). In 2012, the Pediatric Hydroxyurea Phase 3 Clinical Trial (BABY HUG), registered with the National Institutes of Health (Jackson et al., 2020), was a Phase 3 multicenter, randomized, double-blind, placebo-controlled clinical trial of HU in infants (beginning at 9-18 months of age) who were living with sickle cell anemia. This study showed that HU was not associated with an increased risk of bacteremia or serious infection and was generally well tolerated by very young children living with sickle cell anemia. The findings of this study were recently reconfirmed by a 2019 study looking at children aged 5-12 months (Schuchard et al., 2019).

However, despite the strong NHLBI recommendations, uptake has been inconsistent and below recommended levels. One recent study showed the proportion of SCD visits that included new or continued HU prescriptions increased from less than or equal to 8 percent before 2009 to just 33 percent in 2015 to 2017 (Su et al., 2019). In a recent review completed by Centers for Medicare and Medicaid Services (Center for Medicaid and CHIP Services Division of Quality and Health Outcomes, 2020), among CMS pediatric recipients living with SCD (≥21months, ≤ 20 years), 63 percent had no days of HU use in 2017, 21 percent had 1-180 days, and 16 percent had 181-365 days. Among CMS adult recipients (≥21 years, ≤75 years), 65 percent had no days of HU use in 2017, 25 percent had 1-180 days, and 10 percent had 181-365 days.

Possible Factors for Low National HU Rates

- Some providers do not understand or doubt the efficacy of treatment, warranting further education. Studies have shown providers, especially those treating adults, cited this as a reason for not prescribing HU (Brandow et al., 2010; Imegi, 2016; Lanzkron et al., 2008; Zumberg et al., 2005). In one survey conducted with community-based hematologist/oncologists in two southeastern states (Zumberg et al., 2005), 4% of respondents indicated that doubts about the efficacy of HU were "very important" in their decision not to prescribe HU, while 36% rated this variable as "important."
- Provider belief that patient adherence to medications may be low (Brandow & Panepinto, 2010)
- Provider concern about unsubstantiated side effects (e.g., risk of cancer) (Brandow & Panepinto, 2010; Imegi, 2016)
- Lack of patient confidence in HU and need for improving shared decision-making between patient/family/caregiver and provider. Some

patients are concerned about the side effects and safety of HU. Through active engagement and shared decision-making, doctors can provide additional information to address questions so the patient can make an informed decision (Creary et al., 2015; Crosby et al., 2015; Jabour et al., 2019; Sinha et al., 2018).

- Logistical concerns related to social situation may impact patient/caregiver ability to attend follow-up visits for medication monitoring (Loo et al., 2021; National Institute for Children's Health Quality, 2020), including a lack of transportation, work- and school-related conflicts, and distance from the clinic (Cronin et al., 2018).
- The costs of the medication may not be covered by patient's insurance (Treadwell et al., 2020).

Possible Contributing Program Factors for Increased HU Use

RCCs are familiar with and support national guidelines, reporting that some providers begin talking to families about the importance of this drug when the children are as young as 2 months. By the age of 9 months, many families have had the opportunity to think about this treatment option and ask questions. In addition, RCCs have clinical systems in places which assist in making sure that children who are seen by Program providers are started on HU by their first birthday, as recommended by the NHLBI. The RCC collective work has shown that attaining high HU utilization is possible. The success seen in this program shows what can be accomplished through sustained commitment (Karkoska et al., 2021).

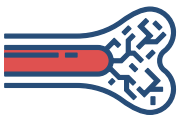
The following are additional SCD medications and therapies. Initial Program data showed a modest upward trend of using other disease-modifying treatments for adults, such as the following medications. Continued tracking of specific types of treatments will be needed to understand if use is sustained or increased.

L-glutamine (**Endari**) was approved in 2017. L-glutamine, appropriate for people 5 years of age and older, has been shown to reduce pain episodes requiring hospitalizations, as well as acute chest syndrome, compared to placebo (Agrawal et al., 2014).

Crizanlizumab (Adakveo), a monoclonal antibody developed by Novartis, was approved by the FDA in 2019 (U.S. Food and Drug Administration, 2019). Crizanlizumab, which is permitted for people 16 years of age and older, helps reduce the frequency of general pain episodes, also known as vaso-occlusive pain episodes (VOCs). This is important as these episodes can escalate to life-threatening conditions and are a major cause of additional costs, such as hospitalization (U.S. Food and Drug Administration, 2019).

Voxelotor (Oxbryta) was granted accelerated approval by the FDA in 2019 for the treatment of SCD in patients 12 years of age and older. This medication can help reduce strokes by increasing hemoglobin levels (HemOnc Today, 2018).

> "On the pediatric side, I'm able to talk to families about the fact that I see patients who are now in their 20s, 30s, 40s, with end-stage organ damage, who either were not on hydroxyurea when they were younger, or were intermittently compliant, and now there's organ damage that can't be repaired. We have the ability to carry that information back to the pediatric patients and talk to families about their realistic expectation of unmodified sickle cell disease. With a family who's been resistant to modifying therapy, we discuss what it looks like, how to weigh the risks of therapies now versus the risks of this disease causing end-stage organ damage later."
>
> MD, Pediatric Hematology-Oncology



Bone Marrow Transplant

At present, the only established cure for SCD is a bone marrow transplant (BMT); most SCD patients are clinically eligible for this procedure. While there are strong rates of success, this procedure is medically complex and potentially fatal. For this procedure, the patient's own marrow is eliminated with chemotherapy followed by a post-procedure recovery period of several weeks to months. In patients, mostly children, who have a matched **allogeneic bone marrow transplantation**, clinicians have seen a 95% success rate. However, BMT is not currently an option for most patients due to the low chance of finding a suitable match for donor bone marrow and costly out-of-pocket hospital expenses for the procedure. The result is that between 1984, when this cure was developed, and the most recent report, only 1,200 U.S. patients with SCD had received a transplant (Bhatia & Sheth, 2015). While this treatment remains out of reach for most patients, given the barriers described, it is currently less expensive and more readily available than experimental gene therapies (Tisdale, 2019).

Gene Therapy

Gene therapy is an emerging field but advancements in gene therapies bring renewed hope for a cure. While some RCCs may have participated in research in this field, no official data were collected on this work during the Program. Given the potential importance of developments, brief information about this therapy is provided here. Based on years of groundbreaking foundational work, the first successful gene-editing procedures were completed in 2020. Clustered Regularly Interspaced Short Palindromic Repeats (CRISPR) Therapeutics has been researching CTX001, an experimental gene-editing cell therapy (Carvalho, 2020; Silva, 2020), and recent successful gene therapy interventions in five patients are giving hope for a cure. Many experts believe that CRISPR therapy has the potential to become widespread during the next 10 years. But hope must be tempered with reality, understanding that more work needs to be done to ensure that these treatments are safe and financially accessible to people living with SCD (Ozuah, 2021).

Disease Prevention

Immunizations

Keeping SCD patients up to date with immunizations is critical and the NHLBI guidelines outline specific time intervals for pneumococcal vaccination. However, national data show that vaccination rates vary widely, both by age and vaccine type (National Academies of Sciences Engineering and Medicine, 2020). While three-quarters or more of patients living with SCD nationally have received at least one of two recommended pneumococcal vaccines, findings show a lower range of patients have received both (30-52 percent). Influenza vaccination ranges from 30-82 percent for pediatric patients and 12-61 percent for adult patients, and only 17-24 percent of patients received the meningococcal vaccine. While there was strong support of the use of this preventive measure, many RCC experienced several barriers to data collection which are described in the data methods appendix.

Screening

Transcranial Doppler

A Transcranial Doppler (TCD) is an important screening that assesses risk of stroke in SCD patients. An **overt stroke** results in apparent neurological deficits such as weakness in an arm or speech problems (Mrkobrada et al., 2019). A **silent stroke** does not cause any noticeable symptoms and can only be seen on brain scans; recurrent ones can cause significant permanent damage. Without prevention, approximately 10 percent of children with SCD by age 20 (Ohene-Frempong et al., 1998) and 24 percent of patients by age 45 will have a stroke (Zétola, 2012). While this is a potential life-saving screening procedure, uptake is low, with one study finding only approximately 45 percent of eligible children being screened (Raphael et al., 2008). Other studies have demonstrated variability in TCD rates. For example, 25 percent of patients aged 2-5 years received at least one screening during a 14-month retrospective cohort study (Bundy et al., 2016), while another study found that 68 percent of 338 publicly-insured children living with SCD (Eckrich et al., 2013) had a TCD during their study period.

The Program has identified the following barriers which may have impeded access to care:

- Limited physical space, staff, or equipment to conduct TCDs
- Limited radiology staff trained to conduct TCD screens

- Providers misunderstanding age cutoff for use of TCDs based on out-of-date information
- Patient distance to clinics with equipment (To address this, some practitioners took TCD machines "on the road," especially to reach remote and rural areas)
- The need for more information about this procedure to address patients' and families' questions and concerns that impact test completion

Pain Management

Pain management of SCD patients is important, but complex.

Provider Resistance to Caring for SCD Pain Crises

People with SCD who present in pain to emergency departments (ED) and other clinical settings need immediate attention. However, research continues to show that provider bias remains problematic (Edwards-Maddox, 2021). Several providers interviewed from the Program concurred noting that some resistance to care for people living with SCD is due to disease stigma, lack of knowledge or comfort treating this population, institutional and/or provider bias, and overt racism. Whatever the reason, this translates to less care. For example, one site called 15 pain specialists in a metropolitan area. None of the pain specialists were willing to see the patient, stating it was "because they have sickle cell disease." From subtle to overt action, providers in the Program perceived inequities and recognized that people living with SCD are held to a different standard of behavior than other patients, such as those with cancer or cystic fibrosis, who may garner more sympathy about their illness. Alternatively, RCCs spoke of how, during this current climate of social/civil unrest, there is renewed hope to close the gap in health disparities by eliminating overt and unconscious bias in the care of this population, particularly in this important dimension of care.

Studies have documented patient dissatisfaction of services received in an ED. One recent survey of people with SCD discussed interactions with ED providers. More than half of patients reported that they did not receive ED care quickly, with wait times exceeding one hour. Additionally, 46 percent and 35 percent also reported feeling that ED physicians and nurses, respectively, did not care about them (Linton et al., 2020). Patients are often assumed to be drug-seeking, which may be a contributing factor to why patients avoid seeking care during pain crises and experience delays in pain medication dispersal (Jenerette & Brewer, 2010; National Academies of Sciences Engineering and Medicine, 2020; Shapiro et al., 1997).

Attention to how providers approach pain management and the need for informed, quality care continues. It is important for providers to watch for, diagnose and address the complications of recurrent and chronic opioid use, across the board. However, specialists remain troubled by incidents of unwarranted restricted opioid treatment (Osunkwo et al., 2020). As patients encounter a pain crisis, they are often forced to navigate stressful ED visits, requiring them to convince emergency and other medical staff that they are not drug-seeking and that their pain is real and needs immediate attention. This prolongs pain episodes, and studies show this is unwarranted. In a recent retrospective observational study (Ballas et al., 2018), authors sought to describe opioid utilization in people living with SCD within the context of healthcare utilization, and the U.S. opioid epidemic among a published broader U.S. population. The authors found that opioid use has remained stable among SCD patients over time, even as opioid use has risen in the U.S. generally. Each patient needs a tailored approach to care and to receive the level of pain control needed. Optimizing this treatment need can be challenging but it is an essential part of SCD care.

> "I've had patients call a hematology practice and say, 'I'd like to make an appointment,' and they say, 'What's your diagnosis?' And they say, 'Sickle cell,' they say, 'We don't see patients with sickle cell.' So... what? You don't see patients? You're a hematology practice. You see hemophilia. You see problems with thrombosis. But you don't see sickle cell? I think that's a huge issue and, in a lot of ways, I agree with my colleague who said it feels like the knee is on the neck of individuals living with sickle cell disease..."
>
> MD, CEO and Medical Director, CBO

Section 3 | National Heart, Lung, and Blood Institute Recommendations and Guidelines

The NHLBI Evidence-Based Management of Sickle Cell Disease (SCD) Expert Panel Report (National Heart, Lung, and Blood Institute, 2014) is recognized as a standard, primary source of information that outlines national guidelines for improving care — both recommendations and consensus panel guidance. This NHLBI report was developed by an expert panel comprised of healthcare professionals with expertise in family medicine, general internal medicine, adult and pediatric hematology, psychiatry, transfusion medicine, obstetrics and gynecology, emergency department nursing, and evidence-based medicine. The purpose of this guide was to provide clinicians, mainly primary care providers, with a digital resource of expert, evidence-based treatment recommendations to guide improvement work on a national scale. The report provides specific recommendations for:

- Comprehensive health maintenance throughout the lifespan
- Effective management of pain episodes and common complications
- Appropriate use of HU and blood transfusions

In the long-term, the use of these guidelines will assist in achieving the collective goal of improving health outcomes and quality of life for people living with SCD. However, according to the Office of Minority Health, only about 1 in 4 people living with SCD receive the standard of care outlined in current guidelines (Office of Minority Health, 2020).

This NHLBI report formed the foundation for many of the targeted quality measures for the Program and continues to be a vital reference for clinicians and sites that take care of people living with SCD. Below are select NHLBI guidelines and/or consensus panel guidance for areas of RCC work completed during this Program:

- HU
- Immunizations
- TCD
- Pain management

The full report can be found at here.

Hydroxyurea

As part of their review process, the NHLBI expert panel examined more than 400 studies conducted between 2007-10 and a prior published NIH Consensus Conference on Hydroxyurea document for studies conducted before that time. This includes the Multicenter Study of Hydroxyurea (MSH) in patients (1992-2008), which was a randomized, double blind, placebo-controlled trial involving 299 adults living with SCD who had experienced three or more **vaso-occlusive crises** (**VOCs**) in the previous year. This trial demonstrated that those on HU treatment had reduced frequency of painful episodes, ACS events, and the need for red blood cell transfusions and hospitalizations. The FDA used this study as the evidence to approve HU for treatment in patients with SCD unless otherwise advised by their doctors not to take it.

HU NHLBI Recommendations:

- Educate all patients with SCA and their family members about hydroxyurea therapy. (See consensus treatment protocol on page 145). (Consensus–Panel Expertise)
- In adults with SCA who have three or more sickle cell-associated moderate to severe pain crises in a 12-month period, treat with hydroxyurea. (Strong Recommendation, High-Quality Evidence)
- In adults with SCA who have sickle cell-associated pain that interferes with daily activities and quality of life, treat with hydroxyurea. (Strong Recommendation, Moderate-Quality Evidence)
- In adults with SCA who have a history of severe/recurrent ACS, treat with hydroxyurea.* (Strong Recommendation, Moderate-Quality Evidence)
- In adults with SCA who have severe symptomatic chronic anemia that interferes with daily activities or quality of life, treat with hydroxyurea. (Strong Recommendation, Moderate-Quality Evidence)
- In infants 9 months of age and older, children, and adolescents with SCA, offer treatment with hydroxyurea regardless of clinical severity to reduce SCD-related complications (e.g., pain, dactylitis, ACS, anemia). (Strong Recommendation, High-Quality Evidence for ages 9–42 months; Moderate Recommendation, Moderate-Quality Evidence for children >42 months and adolescents). Note: The panel intentionally used the term "offer" realizing that patients' values and preferences may differ particularly considering treatment burden (e.g., laboratory monitoring,

office visits), availability of drug in a liquid form, and cost. Therefore, the panel strongly encourages shared decision-making and discussion of hydroxyurea therapy with all patients.

- In adults and children with SCD who have chronic kidney disease and are taking erythropoietin, hydroxyurea therapy can be added to improve anemia. (Weak Recommendation, Low-Quality Evidence)
- In females who are pregnant or breastfeeding, discontinue hydroxyurea therapy. (Moderate Recommendation, Very Low-Quality Evidence)
- To ensure proper use of hydroxyurea and maximize benefits and safety, use an established prescribing and monitoring protocol. (Strong Recommendation, High-Quality Evidence)
- In people with HbSβ+-thalassemia or HbSC who have recurrent sickle cell-associated pain that interferes with daily activities or quality of life, consult a sickle cell expert for consideration of hydroxyurea therapy. (Moderate Recommendation, Low-Quality Evidence)
- In people not demonstrating a clinical response to appropriate doses and duration of hydroxyurea therapy, consult a sickle cell expert. (Moderate Recommendation, Very Low-Quality Evidence)

* For more information, see the ACS section of the "Managing Acute Complications of Sickle Cell Disease" chapter.

Immunizations

The Advisory Committee on Immunization Practices (ACIP) reviews the evidence for each immunization it recommends. The expert panel based its recommendations on those made by the ACIP.

Immunization NHLBI Recommendations:

- All people living with SCD should receive immunizations according to the CDC's Advisory Committee on Immunization Practices (ACIP) harmonized immunization schedule 1 unless they have a personal contraindication as noted in the ACIP schedule.
- Because of their increased susceptibility to invasive pneumococcal disease, all infants with SCD should receive the complete series of the 13-valent conjugate pneumococcal vaccine series beginning shortly after birth and the 23-valent pneumococcal polysaccharide vaccine at age 2 years, with a second dose at age 5 years.

Transcranial Doppler

The NHLBI reviewed two randomized, controlled trials and 50 observational studies in making their recommendations for TCD. The Stroke Prevention Trial in Sickle Cell Anemia (STOP trial) demonstrated a 92 percent decrease in the rate of stroke in children with an abnormal TCD when treated with monthly red blood cell transfusions compared to observation alone (Adams et al., 1998).

TCD NHLBI Recommendation:

- In children with SCD, screen annually with TCD according to methods employed in the STOP studies, beginning at age 2 and continuing until at least age 16.

Pain Management

Pain crises are a hallmark of SCD. Based on a review of several studies, the NHLBI report has several robust clinical recommendations regarding pain management.

Pain Management NHLBI Guidelines:

1. Determine the cause and type of SCD-related chronic pain. This includes chronic pain with objective signs such as avascular necrosis (AVN) and leg ulcers, and chronic pain without objective signs due to neuroplasticity of the peripheral or central nervous system. (Consensus–Adapted)
2. Use a combination of the patient's response to treatment — including pain relief, side effects, and functional outcomes — to guide the long-term use of opioids. (Consensus–Adapted)
3. Encourage people to use deep tissue/deep pressure massage therapy, muscle relaxation therapy, and self-hypnosis as indicated. (Weak Recommendation, Low-Quality Evidence)
4. Use long- and short-acting opioids to manage chronic pain that is not relieved by nonopioids. (Consensus–Adapted)
5. Assess all people with SCD for chronic pain annually or more often as needed. This assessment should include descriptors of the pain; its severity on a numerical scale; its location; factors that precipitate or relieve it, including biopsychosocial factors; and its effect on the patient's mood, activity, employment, quality of life, and vital signs. (Consensus–Adapted)

6. Use a partnership agreement leading to a written, individualized treatment plan (to include risks, benefits, and side effects) with the patient if long-term opioids are indicated. The partnership agreement should list the patient's rights and responsibilities, and the treatment plan should list the type, amount, and route of administration of the opioid in question, including random drug urine testing. (Consensus–Adapted)
7. Appoint one physician or other clinician to write the biweekly to monthly prescriptions for long-term opioids. Refills without seeing the patient should be kept to a minimum, and people on chronic opioid therapy must be evaluated in person every 2–3 months. (Consensus–Adapted)
8. Document all encounters with a patient, including medical history, physical exam, diagnosis, plan of management, type and amount of opioids prescribed and their side effects, if any, and lab data as needed. (Consensus–Adapted)
9. Encourage people receiving opioids to increase their fluid intake, maintain dietary fiber intake per the current dietary fiber recommendations, and to use stool softeners and bowel stimulant laxatives such as senna and/or docusate as needed. (Consensus–Adapted)
10. Believe the patient's report of pain and optimize therapeutic outcomes to achieve adequate pain relief and improve the patient's quality of life. (Consensus–Adapted)
11. Refer patients for evaluation by a mental health professional such as a psychiatrist, social worker, or addiction specialist as needed. (Consensus–Adapted)
12. Assess all people for other types of non-SCD related chronic pain, including postoperative pain, pain due to trauma, pain due to therapy, iatrogenic pain, and pain due to co-morbid conditions. (Consensus–Adapted)

Section 4 | History of Federal Activities Supporting Sickle Cell Disease

The National Heart, Lung, and Blood Institute (NHLBI), part of the National Institutes of Health (NIH), has funded sickle cell disease (SCD) research since 1948, when NHLBI was founded as the National Heart Institute. In 1972, Congress passed the National Sickle Cell Disease Control Act, the first time that the U.S. government formally acknowledged and authorized funds to establish education, information, screening, testing, counseling, research, and treatment programs specifically for SCD. During that time, the NIH oversaw comprehensive SCD research, treatment centers were established, and screening for SCD and education clinics were initiated under the Health Services Administration (HSA), which became Health Resources and Services Administration (HRSA). The following decade (1972-1982) brought advances in all areas relating to SCD, including research, treatment, development of teaching and educational materials, public awareness campaigns, community participation, and community and patient involvement in program development. Continued attention, supported by ongoing legislation, maintained funding mechanisms for programs. This spotlight assisted in building relationships among groups interested in SCD and those dedicated to other genetic diseases.

Universal newborn screening (NBS) for SCD (SCDNBSP) is an NBS program addressing SCD screening. It was established in the late 1980s. This was essential to push early evidence-based treatment protocols for newborns, including vaccinations and prophylactic antibiotic use to prevent infections. Early screening programs have been critical to ensuring timely adoption of treatment options that delay or prevent the complications associated with SCD. Congress established a set-aside for sickle cell in § 501(a) (2) of the Social Security Act as amended (42 U.S.C. 701(a) (2)). HRSA uses this appropriation to support the Sickle Cell Disease Newborn Screening Program (SCDNBS). For individuals identified with SCD and SCT through universal newborn screening, this program improves care and follow-up by supporting efforts of community-based organizations on SCD education and service coordination. The SCDNBSP is separate from the Program described in this report.

Section 5 | Provider Support Strategies

The Program's network of RCCs is a unique national partnership that provided resources and a framework as well as a set of shared priorities. Partners in the network — providers, patients, caregivers, community-based organizations (CBOs), participating sites, Regional Coordinating Centers (RCCs), and the National Coordinating Center (NCC) — pursued a common goal to improve health and healthcare for people living with SCD. The network shared ongoing innovation, learning, and best practices.

Increasing Strength with Diverse Partnerships

People living with SCD, providers, and systems often feel siloed — efforts to improve care, while potentially connected within a locale or institution, are not consistently coordinated regionally or nationally. The partnerships supported through this funding were essential in breaking down silos, both within regions and across the nation. By bringing together diverse stakeholders, the Program encouraged sites to leverage partnerships with hospital administration and pursue bi-directional state and RCC partnerships, including public health stakeholders, to improve SCD care.

The network framework also developed stronger partnerships between clinical care and communities. Community-based organizations (CBOs) were formally involved in all RCCs' networks, collaborating closely with participating sites. RCCs worked with individual advocates, community health workers, patients, their families, and caregivers as they developed and refined programs and outreach to meet the needs of people living with SCD.

Networking and Collaboration on a National Scale

RCCs reported that the Program's coordinated collaborative approach was important in highlighting areas for improvement, e.g., Transcranial Doppler (TCD) screening numbers. Collecting RCC data helped provide a national picture. During regional meetings, sites shared work areas, successes and challenges. This structure allowed for accountability of the work and fostered a sense of camaraderie, partnership, and trust that enabled RCCs to address issues together and make regional improvements.

Trainings for providers, health educators, and CBOs, such as conducting provider-to-provider education and the use of Project ECHO® telementoring programs, were employed to support collaboration. These types of activities helped the network make significant progress toward improving provider competence and confidence in the ability to care for people living with SCD.

The Need to Increase the Number of Well-Trained, Interested Providers

One area of Program work was to improve provider knowledge so that more providers can take care of the SCD population. Increasing provider knowledge had the dual intent of building current provider capacity and engaging providers to become qualified clinicians for the SCD population.

Current Program providers emphasized the need to build a pipeline of providers who are willing and adequately prepared to care for people living with SCD. And while both the pediatric and adult sides of care have seen shortfalls, there has been greater impact for the adult population.

> "That's part of our rounding, too, when we're working with the residents and the attendants with in-patient, we do a lot of teaching on sickle cell. Again, the focus is on trying to increase understanding of how to take care of [people with] sickle cell disease. But even with that said, patients still say, 'There's not enough people, not enough providers, not enough hematologists, to take care of me.'"
>
> APRN, Family Nurse Practitioner

Current Program providers emphasized the need to build a pipeline of providers who are willing and adequately prepared to care for people living with SCD. And while both the pediatric and adult sides of care have seen shortfalls, there has been greater impact for the adult population.

Recruiting providers to specialize in SCD care is challenging. In a recent study (Marshall et al., 2018) looking at factors that lead hematology-oncology fellows to pursue (or not to pursue) careers in **nonmalignant hematology** — SCD is a condition that falls under this care — the authors looked at time spent in different areas of focus. Fellows spent a mean of:

- 52.1 percent of their time in solid tumor oncology
- 37.5 percent in hematologic malignancies
- 10 percent in nonmalignant hematology
- Only 1 percent spent more than 50 percent of time practicing nonmalignant hematology.

The authors found that clinical experience during fellowship was one of the factors most significantly affecting choice of patient population. Increased exposure/access to role models and mentors and opportunities for better career growth/advancement were the top two factors that could have most significantly influenced a decision to spend more time in nonmalignant hematology. Continued mentorship by current specialists and support of direct clinical practice with populations could have a positive influence on future career and pathway choices for trainees.

> We're trying to create [an adult SCD program] in a true comprehensive model where [patients] are seen, and that's hard because the adult hematologists are pushed toward oncology and primary care providers are short on time and staff.
>
> MD, PhD, Pediatric Hematology-Oncology

The lack of providers, particularly hematologists, who are available to care for people living with SCD continues to be a national challenge and affected the care of the RCC sites directly.

Severely under-resourced locations often do not have the staffing or the means to meet the challenges of serving their patients. More well-resourced locations reported being able to increase hematology care by adding physicians and nurse practitioners who specialize in SCD care or to expand other roles to include attention to SCD care, such as advocates, care coordinators, psychologists, and social workers. While consistency in provider care is important in building trusting relationships with patients, having too few providers leads to inadequate staffing and brings the risk of burnout for an already limited pool of providers. Aptly put by one provider, "I could pretty much do everything if I had 40 hours in a day." This concern is so significant, the RCCs have specifically recommended that future iterations of this work include multiple strategies to build a pipeline of providers. Additionally, providers who serve less populated areas or do not work within a structure that includes access to many specialists often find themselves solely responsible for people living with SCD seeking care. These work environments can mean both serving alone and covering large geographic areas, which brings additional challenges. Building and training next generation providers for less populated areas is also needed.

> It's just... nobody seems to care. We have 600 adult patients and a physician assistant who is just coming to work every day, struggling.
>
> MD, CEO and Medical Director of CBO

Caring for people living with SCD is relationship-oriented, often spanning the patient's lifetime — from infancy through adulthood — with providers often caring for the patients' children and other family members. This type of relationship often requires trust built over time. Many of the RCCs and participating sites have extensive experience taking care of people living with SCD. State leads often reported having 20 or more years of experience specifically in SCD care and discussed the importance of their nursing, social work, and allied health teams' years of experience in caring for this population. This lifespan work is important in offering comprehensive care and training and retaining providers is key.

Knowledge-Sharing

Knowledge-sharing among trusted colleagues is an effective way of increasing information. The following section describes the ways RCCs, states, and participating sites shared information and expertise during this Program.

Provider-to-Provider Communication

RCCs conducted provider-to-provider education, formally and informally, to increase capacity to care for this population. All encounters assisted in improving the care of people living with SCD, whether or not they were captured by formal data methods. For example, RCCs reached out to federally qualified health centers to host "lunch and learns" to discuss important clinical and psychosocial issues that impact the lives of people living with SCD and maintained connections to medical resident training programs to connect with younger providers who may be interested in focusing on or expanding their practice to include improving outcomes for people living with SCD. Providers in the Program conducted grand rounds; mentored junior faculty by linking the young physicians with opportunities to directly care for this patient population; initiated and attended health fairs to conduct direct outreach and engagement; and built bridges with local PCPs and ED physicians.

> "Since we don't have a lot of admissions, we had the problem with nurses and other caregivers in the hospital or the ED not continually being educated about them. [The nurse and] our research person and Advance Practice Provider have done these educational sessions we call 'walking tacos,' since they bring snacks for any available staff. They go once a quarter to each unit in the hospital that would have [SCD] patients, [like the ED]. It's eight or nine different units – they've been able, each quarter, to cover about 80 or 90 people in all those units. It's made a significant difference, not just in how the patients are cared for, but in the attitude people have toward the patients. We have a lot more work to do, but that personal interaction is what has helped."
>
> MD, Medical Director, Hematology-Oncology

Telementoring with Project ECHO®

During the past decade, telementoring has been a highly successful approach to remotely supporting clinicians. RCCs used Project ECHO® to increase provider knowledge and have developed a specific recommendation around the use of telementoring.

Project ECHO® was developed in 2002 by Sanjeev Arora, MD, at the University of New Mexico. Project ECHO® is an innovative telementoring program designed to create virtual communities of learners by bringing together healthcare providers and subject matter experts using videoconferencing to host brief lecture presentations and case-based learning. Fostering an "all learn, all teach" approach, participants engage in the bi-directional virtual knowledge network by sharing clinical challenges and learning from experts and peers. Project ECHO® has been recognized globally as a successful tool for improving patient care outcomes. Being grounded in telementoring puts the focus on provider-to-provider communications in addition to the provider and patient relationship-building.

Project ECHO® follows these four key principles:

1. Subscribes to the disease management model of care that aims to improve quality, while reducing variety of and standardizing best practices

2. Fosters multidisciplinary partnerships that increase access to care and reduce health care costs
3. Engages health care providers to participate in case-based learning under guided practice to provide specialized care to their own patients
4. Utilizes technology to promote face-to-face mentorship and sharing of knowledge and experience by experts and peers without the need for cost-intensive supervision, in-person trainings, or travel

Project ECHO® has published data demonstrating that patients who received care from providers mentored using the Project ECHO® methodology had outcomes as good or better than those treated at specialized referral hospitals.

While Project ECHO® has been successfully used to improve management of chronic conditions (e.g., Hepatitis C), the use and testing with rare chronic conditions such as SCD is new. However, given Project ECHO®'s strong track record, RCCs were asked to use the Project ECHO® model or similar models to provide education and mentoring to providers to expand reach, especially to geographic locations where providers are isolated (either due to lack of specialists or a rural location) and could benefit from a network of established SCD providers. Sessions were open to all types of clinicians and care team members, with the goal of enabling PCPs to have ready access to SCD specialists. The structure of this Program provided a prime opportunity to initiate education and training models for SCD care.

With the use of regional provider expertise, the lead RCC and/or states were encouraged to coordinate, implement, and evaluate regularly scheduled telementoring/telemedicine programs and clinics. Within the funding cycle, the regions and states carefully worked to refine programming.

Addressing Provider Comfort with Treating People Living with SCD

Understanding guidelines for how to take care of people living with SCD is important, and this Program supported RCCs to develop and offer several options. At the same time, RCC experience has shown the ability to take care of this population goes beyond knowing the guidelines. Provider comfort is an important factor in being able to appropriately care for this population. For providers who have small numbers of patients, such as PCPs or specialists who do not regularly take care of patients (e.g., ED providers, cardiologists, pulmonologists), having the bandwidth to devote time or resources to keep up with current care guidelines for this population with specific needs can be daunting (National Academies of Sciences Engineering and Medicine, 2020). This contributes to a circular pattern of discomfort, which may be interpreted as an unwillingness to be involved with the care. Each educational session is an opportunity to improve providers' comfort with treating people living with SCD.

Section 6 | High-Quality Care

Achieving optimal health outcomes for people living with SCD depends on delivering comprehensive, coordinated care features of a **medical home** model. RCCs report that when care is coordinated, they can improve access and quality that, in turn, improve outcomes. All RCCs completed activities in care coordination. Also, for children and young adults, continuous care includes ensuring successful care transition between pediatric and adult care providers.

> We're fortunate that the hospital has decided to support care of adult patients. I was hired to have that focus – clinically helping the adult nonmalignant hematology patients. The hospital recognized this need because they have been caring for these patients and seeing that when they reach adult age, [there is] no other medical home available that had the expertise to become the primary medical home to care for patients with sickle cell disease.
>
> MD, Pediatric Hematology/ Oncology

Comprehensive, Coordinated, and Continuous Care

Comprehensive Care: Addressing Psychosocial Needs

Comprehensive care for this population includes attention to both physical and psychosocial needs. While much clinical knowledge has been gained about SCD, there is a paucity of psychosocial information, such as the impact of the condition on a person's mental health and daily functioning, documentation on years of life lost, other areas of morbidity, and the social stigma of SCD.

This Report has provided information on treating physical issues related to SCD. But providers relayed the need for greater attention to psychosocial issues and social realities for patients. For example, one site pointed out the challenge of planning care transition for young adults who may not have housing, or discussing the importance of eating a balanced meal, taking HU, managing a complicated condition, or not seeing follow-through with children whose home life may not be stable.

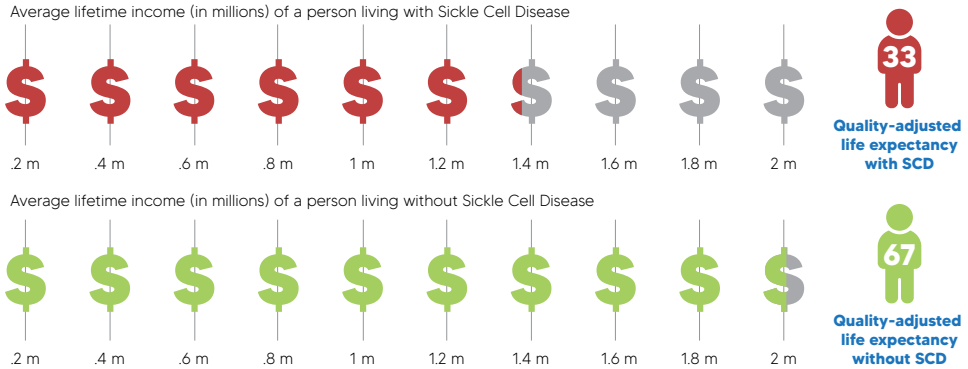
Sites used both traditional methods of care and service and innovative programming to address these needs, including patient-centered strategies like self-hypnosis, biofeedback, yoga, and mindfulness; some implemented new protocols, including having children with SCD seen by a pediatric psychologist early in care; others dedicated social workers, SCD health advocates, and pediatric neuropsychologists to address existing and emerging needs. Using regular mental health assessments (PHQ-9, a measurement for depression, and GAD-7, a measurement for anxiety disorders), some sites discovered that patients who have pain issues often have emotional issues and distress that may be exacerbating their pain. To address this, they focused on pain and

> We are also learning about mental and behavioral health issues. This is something families are openly and loudly talking about. Depression and other concerns across the spectrum, from suicide to substance abuse and running away from home. We are running behind and trying to catch up to address these family issues.
>
> President and CEO, CBO

mental health issues concurrently. RCCs all noted an increased demand for mental health services during the pandemic and created programming to meet the need and used telehealth for behavioral health appointments.

There are geographic, economic, and sociocultural barriers that may impede access to quality care. These psychosocial factors can impact quality of life for a person with SCD, which is important as expected lifespan for people with SCD has increased. In recent history, many people with SCD died during childhood. In the early 1970s, the average lifespan of a person living with SCD was 14 years of age (National Heart Lung and Blood Institute, 2010). Now, an estimated 94 percent of children living with SCD will live to adulthood (Agency for Healthcare Research and Quality, 2020), and half of patients with SCD will live beyond 50 years of age (Platt et al., 1994). Though there have been life expectancy gains, a recent article (Lubeck et al., 2019) that examined life expectancy, quality-adjusted life expectancy (Weinstein & Stason, 1977), and income differences between people living with SCD to those who do not have the condition showed substantial differences. Using a simulated cohort model, the authors found the quality-adjusted life expectancy was 33 years for a person living with SCD compared to 67 years for a person living without. Income differences between people living with SCD and those living without SCD was substantial, with those living with SCD making $1,227,000 during a lifetime and the comparison group making $1,922,000 — a lost income of $695,000 due to decreased life expectancy. Continued work is needed to better understand and address these areas and recommendations related to mental health and other psychosocial support are provided in this report.

Figure 1. Quality of Life Impacts for People Living with SCD



Lubeck, D., Agodoa, I., Bhakta, N., Danese, M., Pappu, K., Howard, R., Gleeson, M., Halperin, M., & Lanzkron, S. (2019). Estimated life expectancy and income of patients with sickle cell disease compared with those without sickle cell disease. *JAMA Network Open, 2*(11), e1915374-e1915374. https://doi.org/10.1001/jamanetworkopen.2019.15374

Coordinated Care

The experience of the RCCs in the Program recommends that an SCD expert, when available, coordinate care. Furthermore, RCCs identified the following areas where better coordination and communication could improve access to quality care:

- Within individual healthcare systems
- Between community sites and major treatment centers
- Among providers (especially during the time of transition from pediatric to adult care)
- In public health research studies and surveillance

In August 2020, directors of top SCD programs in the United States met to publish the first set of recommendations for establishing adult SCD care centers. The recommendations, published in *Blood Advances*, codify the required components of establishing SCD adult care centers.

> Infants are referred early and the primary care provider (PCP) establishes medical homes in partnership with our specialty services. Then the PCP makes the referral to us for specialty care, and we stay in close communication. This joint care approach allows us to offer hydroxyurea therapy when the babies turn nine months old, which is the recommended standard of care. Since we already have established a relationship with the families, they are more inclined to consider hydroxyurea early. When they turn two years old, implementation of the Transcranial Doppler screening for stroke risk and pneumococcal vaccine provided protection against invasive serious infections.
>
> MD, Pediatric Hematology-Oncology

Requirements include:

- Multidisciplinary, team-based, evidence-guided care that is coordinated throughout the institution
- The SCD center as the recognized authority for managing SCD within the institution
- A physician lead who is considered an SCD specialist
- One or more social workers, a patient coordinator, and dedicated nursing staff
- The ability to offer acute and chronic pain management, transfusion, and access to specialists

Comparing these recommendations with current participating site offerings will be important in ensuring consistent, comprehensive care for SCD patients in the future.

MODEL SCD CLINIC

Substantial time has been spent to create guidance for sites who want to know how to set up a model SCD clinic, which includes many important aspects of comprehensive medical homes. These detailed guidelines are available for reference in Section 16 of this Appendix. It covers factors to consider, and specific components needed for a model SCD clinic. (Kanter et al, 2020)

> "Four half-days a month we have our comprehensive sickle cell clinic. Historically, [the patient] would come see the hematologist, then get their echocardiogram, and then come back again to see the pulmonologist, and then they need their neuropsychiatric testing. It just wouldn't happen, right? The kids didn't get what they needed. We know it's a long day, and most families appreciate it, but the kids will come once a year."
>
> MD, Pediatric Hematology-Oncology

Dedicated Space

Having access to dedicated clinic space for SCD care was not guaranteed at all sites, let alone the ability to collaborate as a team, though this has been shown to be beneficial. One recent study (Pujalte et al., 2020) found that co-locating or grouping care team members for both visual and verbal communications is likely to enhance communication and teamwork, resulting in more efficient care for patients. This physical structure has helped lead to clearer, faster communication between care team members. Another study (Rumball-Smith et al., 2014) examining the specific needs of chronic disease management said that care for people living with SCD is the "challenge of the century" and that co-location of practitioners may improve access to services and equipment that aid chronic disease management.

Continuous Care

Increased Use of Telehealth and Virtual Care

The use of telehealth visits prior to the COVID-19 pandemic and robustly used during the pandemic was one of the innovative ways the Program met the needs of patients to ensure ongoing care.

> "One of the things being picked up in the telemedicine surveys is the closeness of the relationship on that telemedicine visit. Patients felt more comfortable telling you things on the telephone that they never used to tell you about: family problems, the problems going on. They were actually more open to it on the phone than they were in person."
>
> MD, Pediatric Hematology-Oncology

With the pandemic lingering and clinical medicine likely forever changed in some ways, continuing to develop and build capacity for telehealth and address barriers is warranted. While many clinics, especially pediatric units, have returned to care-as-usual and many providers feel strongly that the best care that they can

give to their SCD patients is in person, all still recognize that telehealth can supplement in-person care for many people with SCD.

Bridging the Gap: Safe Transfer of Care from Pediatric to Adult Medicine
As life expectancy has increased, ensuring the safe and comprehensive transition from pediatric to adult care is essential in providing continuous care. Early adulthood remains a critical period for SCD patients as they transition care — a process that relies on the availability of qualified adult healthcare providers and healthcare facilities that can meet their needs. Research has shown that patients with SCD who live into adulthood experience overall lower access to care and reduced quality of care relative to the pediatric population with SCD (Haywood Jr et al., 2009; Maxwell et al., 1999). Improving transition from pediatric to adult care and optimizing adult care are both needed for people living with SCD.

RCCs noted two outstanding system challenges:

1. While life expectancy has increased for people with SCD, additional support and structure is needed to create a system to ensure consistent transfer from pediatric to adult clinical care. One study (Sobota et al., 2011) showed that only 60 percent of pediatric SCD centers were able to regularly transfer patients with SCD to an adult hematologist that specialized in SCD. The transitional care offered in most clinics is inconsistent, often incomplete, and sometimes not available at all.

> "Initially, we had very few readiness plans, which included all the education that we needed to accomplish before that patient was ready to transition. When we started our QI project, we had very few [with] a readiness plan in place. Now, when we do our chart audits, we consistently have 90 percent and greater of patients who have a transition plan in place [and] have done a readiness survey within six months."
>
> MD; Pediatric Hematology-Oncology

2. The lack of adult care coupled with varying quality transitional care means that the oldest of the patients in pediatric care are in limbo: stuck in adolescence, without proper guidance about how to take charge of their own healthcare but expected to be responsible for directing their complex treatment, including dealing with the bias and discrimination that they face in the ED with a pain crisis. This is particularly important as findings show that young adults living with SCD are more likely than any other age group of patients with SCD to seek care in the ED around the time of transitioning care to an adult provider (National Academies of Sciences Engineering and Medicine, 2020).

The importance of the completion of care transition from pediatric to adult medicine is well documented and the Program has made a specific recommendation on this topic. Yet, major barriers to improvement exist. Faster progress could be made in pediatric-to-adult care transition services with focused staffing, such as a transition coordinator to help oversee transition registries.

> "Some big successes have been the addition of our sickle cell family health advocate and how that has translated into better outcomes for our patients. We could focus on hydroxyurea – our trust building, our acceptance of starting hydroxyurea during infancy has been fantastic. Our rate of patients missing appointments has plummeted."
>
> MD, Pediatric Hematology-Oncology

An Essential Element for Quality SCD Care: Engaging and Partnering with Patients, Families, Caregivers and Community-Based Organizations

Engaging patients, families, and caregivers in their own medical care has shown improved adherence to medication, better self-care, and patient satisfaction. Community-based organizations (CBOs) have an important role in helping patients and their caregivers to engage in their own care, and to serve as partners with clinics, providers, patients, and families who seek to learn more and improve health outcomes.

> "Caring for patients is very relationship-oriented, and building trust is key. Our health equity advocate and community health worker can build trust with a family in four minutes that might take me four hours, four months, four years."
>
> MD; Pediatric Hematology-Oncology

Engaging with Patients and Families to Increase Accessibility of Appointments and Remove Barriers to Care

Missed appointments cause delays in care for patients and are an administrative burden on the healthcare system. Missed appointments for SCD patients may be substantial. In a survey of 542 adult patients and caregivers of pediatric patients with SCD, 87 percent of adults with SCD and 65 percent of caregivers reported missing at least one appointment in the previous 12 months (Cronin et al., 2018). In this study, there were varied reasons for missing an appointment, with financial barriers largely contributing to missed appointments.

Clinics used several strategies, including integrating health advocates, to address fundamental patient barriers to appointments and medical care. RCCs have done work to build patient trust, help with medical costs, provide insurance, and offer culturally sensitive care to increase access.

> "The question they ask patients is, 'What do [you] want out of life?' and 'How can the provider and clinic help [you] plan for and get to that destiny?'"
>
> APRN, Family Nurse Practitioner

> "The clinic was set up to be once every three months. If you miss an appointment, then you miss a lot. I can't really keep the SS (most common type of SCD) patients waiting for six months, especially those who should be taking hydroxyurea and screening or TCDs. If you miss a couple appointments, you miss the year."
>
> MD, Adult Hematology-Oncology

As noted in the main report, not all HU formulations are covered by insurance. It is most readily available as a pill; liquid hydroxyurea, needed for some patients, such as small children, is not easily available or affordable for all, even as of this Report. To ensure availability of HU, some sites have worked with their health system pharmacies to compound (mix) the drug on site. Though the partnership and willingness to do this is essential, so is the money needed to compound, which is often $40-$70 per patient with no Medicaid reimbursement available. Additionally, even for patients who are willing to take the medicine once available, burden in staff time and cost can still fall to the clinics, such as arranging shipping medications to those where transportation is a barrier.

Program clinics have had to be creative to fill needs, such as finding grants and donations to cover costs. Because Medicaid would not pay for compounded HU, one site worked with their pharmacy to make and ship it free as a trial. However, patients or caregivers were required to be at home to receive it. Due to work schedules and housing instability of some patients, this took quite a bit of staff effort. Managing administrative issues like these is taxing and not feasible for all sites but is needed to reduce barriers to use HU.

> People have huge transportation issues. Some of the areas for the outreach clinics are an hour and a half from our main clinic but they may still be 20-30 minutes from where the patient lives. We've also been able to work with Medicaid and philanthropic groups to get some transportation for patients who live 45 minutes or so from our main clinic and need to come there for appointments.
>
> MD, Medical Director, Division of Hematology-Oncology

> I have patients who only speak French, so I need help with the hydroxyurea educational materials. Everybody throughout the region resourced, everybody worked to get it documented. Eventually, that document ended up at the CDC.
>
> MD, Chief of Pediatric Hematology

SCD affects people across the world. Some Program regions have experienced an increase in their immigrant and refugee communities; attending to the needs of this specific group within a participating site's patient population is warranted. To effectively care for these groups of people with SCD, screening, clinical care, and materials must be culturally and linguistically appropriate and sensitive. Developing processes to accurately understand the distinct needs of these communities, including partnering with experts in refugee and immigrant care to ensure quality care, should be pursued. The Program has developed a specific recommendation regarding this topic.

Patient-Centered Materials

Quality care includes accessible patient education materials (literacy, language, culturally appropriate, targeted), so patients can gain knowledge and make informed decisions. The RCCs have developed an array of resources, covering a wide variety of SCD topics. These are categorized in the Compendium of Tools and Resources section of this report. Clinics across the country can access and freely use these resources as needed. The Compendium was updated in 2018 and, prior to that, in 2017. This continuous review and sharing of materials for content, appropriateness, and effectiveness is essential.

Clinicians share accurate information with patients, both verbally, written, and using other mediums, such as video. All methods are important given that some patients have cognitive challenges because of their SCD. RCCs have worked to dispel rumors about established medications and answer questions about new medications, especially addressing, disease-modifying therapies, gene therapy innovations, bone marrow transplant process, contraception, eye care, and a myriad of other topics.

Partnering with SCD CBOs

CBOs are essential in engaging with patients and families. They understand what families need and listen to the patient voice. Through this Program, CBOs and clinics have strengthened their partnerships, which has been instrumental in establishing and maintaining an array of services for people with SCD. Yet, this work is not easy and CBOs discussed some of the challenges.

CBO Funding

Well-established CBOs shared the need for additional staff, with strong executive direction being foundational. But few have funds to employ a full-time director. Without consistent and dedicated leadership, it is difficult to have a program that is organized, efficient, and responsive to patients' and communities' needs. Additionally, raising funds necessary to support the mission is an expertise that is essential to the sustainability of an organization — however, many CBOs cannot afford to hire this additional expertise. So, it is important that the CBO leadership have this experience or can be mentored by someone who does. In some cases, the RCC leads of the program have been instrumental in helping local CBOs apply for funding and mentoring them throughout the process. The CBOs in this Program are committed to helping each other, within and across RCCs. The bigger, longer-established CBOs have mentored smaller ones or those in startup phase. Some larger CBOs also help others with funding, making fundraising efforts work for the collective good of elevating all CBO Program work.

From an administrative perspective, raising money needed to operate a CBO is difficult as there is often steep competition, sometimes with medical clinics and other CBOs, for the small dollars that are available for SCD. This competition may be unnecessary if greater alliances could be formed that capitalize on the strengths of each for funding availability and synergies should be pursued. Some CBOs found success obtaining block grants. These awards are disbursed to a single state or community government agency that has local authority to allocate to a wide range of services. If unrestricted, these funds can help with brick-and-mortar costs or partnering with clinics to help with staff costs as part of a formal partnership.

> "I know nonprofits are nonprofits, but [we] are businesses also. You have to have that capacity to do that kind of work, have the accountability, and all the other things that would be critically important to really be able to sustain yourself."
>
> President and CEO, CBO

> "We're starting a program where we can send hydroxyurea to the families that live a couple hours away. I had a fraternity that donated $1,000 and said, 'We want you to improve the care of sickle cell patients.' I thought, 'What can I do to make this a reality?' So, we're starting a program where we can send HU to the families. I can literally look this guy in the eye and say that I've had several parents who've said, 'I'd take that medicine if you figured out how I could get it.' And so now we can afford to pay for the shipping and handling."
>
> MD, Pediatric Hematology-Oncology

Section 7: National Academies for Sciences, Engineering, and Medicine (NASEM): A Blueprint for Action

The Office of Minority Health at the Office of the Assistant Secretary for Health at the U.S. Department of Health and Human Services (HHS) requested that the National Academies for Sciences, Engineering, and Medicine (NASEM) convene a committee to develop a strategic plan and blueprint to address SCD in the United States. The Committee on *Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action* was established in response to this request. Several members of the Program RCC teams and Oversight Steering Committee were involved with this project, able to provide both their clinical expertise and first-hand experience from the Program during the creation of the NASEM report. As part of the application process for the Program funding, each of the regions was charged with creating regional Action Plans that outlined current available resources and infrastructure. These action plans were important to supporting state SCD programming growth as well as instrumental resources in the creation of this blueprint. Although this Blueprint was not part of this project funding, the NCC strongly recommends reviewing the NASEM report for the research done in these areas and the important perspectives shared. **Without concerted attention and change, forward progress in the care of patients living with SCD will be inherently limited.**

The NASEM ad hoc committee was charged to examine:

- The epidemiology, health outcomes, genetic implications, and societal factors associated with SCD and sickle cell trait (SCT), including serious complications of SCD, such as stroke, kidney and heart problems, acute chest syndrome, and debilitating pain crises;
- Current guidelines and best practices for the care of patients with SCD;
- The economic burden associated with SCD; and
- Current federal, state, and local programs related to SCD and SCT, including screening, monitoring and surveillance, treatment and care programs, research.

The committee offered deep insight into critical areas, including screening, registries, and surveillance topics, along with recommendations on delivering high-quality SCD care with a prepared workforce. In addition, the authors did an extensive look at societal and structural contributors to poor disease outcomes, including racism, implicit bias, and socio-economic barriers. These latter areas are significant in the context of SCD care and require great attention, which was outside of the scope of this program. The strategic plan consists of eight overarching strategies, or "pillars," that support the vision, and seven foundational principles that undergird the strategic plan.

Strategic Vision: Long, healthy, productive lives for those living with sickle cell disease (SCD) and those with sickle cell trait (SCT).

- Establish a national system to collect and link data to characterize the burden of the disease, treatment outcomes, and the needs of those with SCD across the life span
- Establish organized systems of care that ensure both clinical and nonclinical supportive services to all persons living with SCD
- Strengthen the evidence base for interventions and disease management and implement widespread efforts to monitor the quality of SCD care
- Increase the number of qualified health professionals providing SCD care
- Improve SCD awareness and strengthen advocacy efforts through targeted education and strategic partnerships among the U.S. Department of Health and Human Services, health care providers, advocacy groups, community-based organizations, professional associations, and other key stakeholders (e.g., media and state health departments)
- Address barriers to accessing current and pipeline therapies for SCD
- Implement efforts to advance understanding of the full impact of SCT on individuals and society
- Establish and fund a research agenda to inform effective programs and policies across the life span
- Foundational Principles: **Safe · Effective · Patient-centered · Timely · Efficient · Equitable · Ethical**

The NCC highly recommends that Congress review the NASEM report and recommendations along with this report, as both provide valuable insight into the comprehensive nature of the work that is being conducted to improve care provided to people living with SCD and their families and outstanding work still to be done. Findings of the NASEM blueprint and this report often reflect and support each other, as the work done for this condition is carried out by a limited but intensely dedicated group of medical teams, researchers, and stakeholder advocates. The challenges and recommendations conveyed in the blueprint and this report are similar in addressing comprehensive, evidence-based care to help people living with SCD live their best lives.

Section 8 | Acronym List

ACIP	Advisory Committee on Immunization Practices
ACS	Acute Chest Syndrome
APP	Advanced Practice Professional
APRN	Advanced Practice Registered Nurse
ASH	American Society of Hematologists
BMT	Bone marrow transplant
CASCADE	Community Access for Sickle Cell ADult CarE
CBO	Community-Based Organization
CCHMC	Cincinnati Children's Hospital Medical Center
CDC	Center for Disease Control and Prevention
CDE	Common Data Elements
CQIM	Clinical Quality Improvement Measures
DCSHCN	Division for Children with Special Health Care Needs
DISPLACE	Dissemination And Implementation of Stroke Prevention Looking at the Care Environment
DMWG	Data Management Working Group
ECHO	Extension for Community Healthcare Options
ED	Emergency Department
EHR	Electronic Health Record
EMBRACE	Education and Mentoring to BRing Access to CarE
FOA	Funding Opportunity Announcement
GRNDaD	Globin Research Network for Data and Discovery
H.U.G.S.	Hugs, Uncomplicated, Gratifying Support Services
HHS	Health and Human Services
HIPAA	Health Insurance Portability and Accountabilty Act
HOPS	Hydroxyurea Optimization through Precision Study
HRSA	Health Resources and Services Administration
HU	Hydroxyurea
IHTC	Indiana Hemophilia and Thrombosis Center
IT	Information Technology
MCHB	Maternal and Child Health Bureau
NASEM	National Academies for Sciences, Engineering, and Medicine
NCC	National Coordinating Center
NHLBI	National Heart, Blood and Lung Institute
NICHQ	National Institute for Children's Health Quality
OSC	Oversight Steering Committee
PCI	Pneumococcal Immunization
PCP	Primary Care Physician
PI	Principal Investigator
PM	Performance Measures
PSCRC	Pacific Sickle Cell Regional Collaborative
PSPM	Provider Survey for Performance Measures
QI	Quality Improvement
QUAKS	Quality improvement for Urea Adherence in Kids with Sickle Cell Disease Study
RCC	Regional Coordinating Centers
RFP	Request for Proposal
SCA	Sickle Cell Anemia
SCD	Sickle Cell Disease
SCDF	Sickle Cell Disease Foundation
SCDTDRCP	Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program
SCT	Sickle Cell Trait
SINERGe	Sickle Cell Improvement in the Northeast Region through Education
ST3P-UP	Sickle cell Trevor Thompson Transition Study
STOP	Stroke Prevention Trial
STORM	Sickle Treatment and Outcomes Research
TCD	Transcranial Doppler
YAPFAQ	Youth Acute Pain Functional Ability Questionnaire

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Appendix A: RCC Activities

Section 9 | List of Data Tables and Figures

Appendix B: Sickle Cell Disease Overview & Background

Appendix C: Data Methodology

Model Protocol

Quarter 1 QI Data Summary

Overview: Quarter 1 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) spanned January 1 to March 31, 2019[1]. RCCs aggregated and submitted data to CoLab between April 1 and May 15, 2019. Data were intended to be a population-level view of key Quality Improvement metrics for all regions participating in SCDTDRCP (Hydroxyurea use, other disease modifying therapy use, immunization status, Transcranial Doppler Ultrasound screenings, transitions to adult care, and providers participation). Regions collected data for this QI report from Electronic Health Records (EHR) or other medical records (such as manual chart review) at the site level. The sample for each region was intended to reflect the total number of sickle-cell patients seen within the specific quarter. For Quarter 1, three out of five regions submitted data (Northeast, Midwest, and Heartland/Southwest). Among the regions that submitted data, not all sites in the Midwest were able to report on values. All eligible sites from the Heartland/Southwest provided data; it is unclear whether all sites within the Northeast reported values based on the data submitted to CoLab. All sites reported issues accessing data from medical records, particularly for immunizations.

The first page of this report displays the aggregated values for the QI measures across all regions. The following pages of this report provide QI measures and values by region. Notes about annotations are provided where relevant. Guidance to RCCS about inclusion for numerators and denominators is detailed in the MOP. Linked here is the most up-to-date MOP.

Aggregated Values for QI Measures (excluding QI Measure 4, immunizations)

	QI Measure 1: HU Use			QI Measure 2: Disease Modifying Therapy Use			QI Measure 3: Patients with TCD screening in past 15 months[2]			QI Measure 5: Transitions to Adult Care[3]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	1904	3279	58.1	240	1908	12.6	1426	3038	46.9	111	323	34.4	362
Adult	776	1333	58.2	262	760	34.5							

Note: N = Numerator; D= Denominator

1. For one region, data collection was from September 1, 2018 – February 28th, 2019
2. Only for eligible patients between 2 and 16 years of age
3. Only for patients ≥ 14 and < 17 years of age

Aggregated Values for QI Measure 4 (Immunization Status)

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	1579	2756	57.3	1612	2556	63.1	1170	2763	42.3	1573	2775	56.7	1142	2740	41.7	714	2450	29.1	383	2529	15.1
Adult	1128	2461	45.8	847	2495	33.9	907	2495	36.4	905	2495	36.3	719	2493	28.8	184	2461	7.48	255	2461	10.4

Note: N = Numerator; D= Denominator. There may be small variation between sites due to initial confusion about how SCDTDRCP considered patient eligibility for immunizations and sites' accordingly varying classification of up-to-date status. We have clarified this issue with sites and resolved the issue moving forward.

***Vaccine priority for this project**

QI Measures by Region

QI Measure 1a: (Hydroxyurea use among Children/Adolescents) Percent of patients >9-months and <18 years of age prescribed Hydroxyurea

Region	Numerator	Denominator	Percentage
Northeast	643	883	72.8%
Midwest	345	437	78.9%
Heartland/Southwest	916	1959	46.8%

QI Measure 1b: (Hydroxyurea use among Adults) Percent of 18 years and older prescribed Hydroxyurea

Region	Numerator	Denominator	Percentage
Northeast	485	874	55.5%
Midwest	75	108	69.4%
Heartland/Southwest	216	351	20.5%

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents) Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU (Optional)

Region	Numerator	Denominator	Percentage
Northeast	62	275	22.5%
Midwest	54	413	13.1%
Heartland/Southwest	124	1220	10.2%

QI Measure 2b: (Disease modifying therapy use other than HU among Adults) Percent of 18 years and older prescribed disease modifying therapy other than HU (Optional)

Region	Numerator	Denominator	Percentage
Northeast	161	406	39.7%
Midwest	21	104	20.2%
Heartland/Southwest	80	250	32.0%

QI Measure 3: (Transcranial Doppler): Eligible SCD patients between ages of 2 -16, that had a Transcranial Doppler (TCD) screening within the last 15 months

Region	Numerator	Denominator	Percentage
Northeast	393	881	44.6%
Midwest	299	371	80.6%
Heartland/Southwest	734	1786	41.1%

4

QI Measure 4a: (Immunization) Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Note: Northeast region did not ask for denominator per vaccine. To note, this is the correct method. The RCC counted all potential patients in the denominator. Patients who are not eligible (e.g. age ineligible were counted in the numerator as up to date). Midwest corrected their data so that there was only one denominator per vaccine. Corrected data for the Midwest is reported. In addition, the Midwest noted not all sites reported. Heartland/Southwest reports issues accessing data.

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Northeast	598	1493	40.1	754	1493	50.5	448	1493	30.0	525	1493	35.2	316	1493	21.2	38	1493	2.55	119	1493	8.00
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8
Heartland/ Southwest[2]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50

[1.] Up to date [2.] Regions where estimates may not be accurate

QI Measure 4b: (Immunization) Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Note: Heartland/Southwest reports issues accessing data. Midwest states not all sites reported. Midwest corrected their data so that there was only one denominator per vaccine. Corrected data for the Midwest is reported. Northeast stated that they did not ask for denominator per vaccine (though this is the correct method)

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.16
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Heartland/ Southwest[2]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	152	295	51.5	20	263	7.60	13	263	4.94

[1.] Up to date [2.] Regions where estimates may not be accurate

QI Measure 5: (Transitions to Adult Care) Number of patients that have a documented transition education

Region	Numerator	Denominator	Percentage
Northeast	21	110	19.1%
Midwest	9	48	18.8%
Heartland/Southwest	81	165	49.1%

QI Measure 6: (ECHO) Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Count
Overall	362
Northeast	170
Midwest	131
Heartland/Southwest	61

Quarter 2 QI Data Summary

Overview: Quarter 2 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) spanned April 1 to June 30, 2019[1]. RCCs aggregated and submitted data to CoLab between July 1 and August 15, 2019. Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers participation in ECHO

Regions collected data for this QI report from Electronic Health Records (EHR) or other medical records (such as manual chart review) at the site level. The sample for each region was intended to reflect the total number of sickle-cell patients seen within the specific quarter. For Quarter 2, three out of five regions submitted data (Midwest, Southeast and Heartland/Southwest).

The first page of this report displays the aggregated values for the QI measures across regions. The following pages of this report provide QI measures and values by region. Notes about annotations are provided where relevant. Guidance to RCCs about inclusion for numerators and denominators is detailed in the MOP. Linked here is the most up-to-date MOP.

Aggregated Values for Q2 QI Measures (*excluding QI Measure 4, immunizations- see below*)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[2]			QI Measure 5: Transitions to Adult Care[3]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	2832	4631	61.2	382	3027	12.6	1666	3189	52.2	246	553	44.5	161
Adult	1448	2451	59.1	421	2448	17.2							

Note: N = Numerator; D= Denominator

1. Three out of five regions included on April 1 to June 30, 2019 timeframe and are included in the aggregate data.
2. Only for eligible patients between 2 and 16 years of age
3. Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q2 QI Measure 4 (Immunization Status)

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Pediatric	1254	1633	76.8	1371	1633	84.0	1029	1633	63.0	1250	1633	76.5	999	1633	61.2	923	1633	56.5	351	1633	21.5
Adult	286	504	56.7	349	504	69.2	305	504	60.5	272	504	54.0	274	504	54.4	173	504	34.3	78	504	15.5

Note: N = Numerator; D= Denominator. Southeast did not provide vaccination data. Northeast did not provider data for pediatric MenACYW and adult Flu.

***Vaccine priority for this project**

Region Specific Values for QI Measures

QI Measure 1a: (Hydroxyurea use among Children/Adolescents) Percent of patients >9-months and <18 years of age prescribed Hydroxyurea – 6 months preceding June 30, 2019

Region	Q1			Q2		
	Numerator	Denominator	Percentage	Numerator	Denominator	Percentage
Heartland/Southwest	916	1959	46.8	942	1938	48.6
Midwest	345	437	78.9	338	393	86.0
Southeast				1552	2300	67.5

QI Measure 1b: (Hydroxyurea use among Adults) Percent of 18 years and older prescribed Hydroxyurea - 6 months preceding June 30, 2019

Region	Q1			Q2		
	Numerator	Denominator	Percentage	Numerator	Denominator	Percentage
Heartland/Southwest	216	351	61.5	201	357	56.3
Midwest	75	108	69.4	90	119	75.6
Southeast				1157	1975	58.6

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents) Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU (Optional) - 6 months preceding June 30, 2019

Region	Q1			Q2		
	Numerator	Denominator	Percentage	Numerator	Denominator	Percentage
Heartland/Southwest	124	1220	10.2	120	1085	11.1
Midwest	54	413	13.1	58	540	10.7
Southeast				204	1402	14.6

QI Measure 2b: (Disease modifying therapy use other than HU among Adults) Percent of 18 years and older prescribed disease modifying therapy other than HU (Optional) - 6 months preceding June 30, 2019

Region	Q1			Q2		
	Numerator	Denominator	Percentage	Numerator	Denominator	Percentage
Heartland/Southwest	80	250	32.0	91	237	38.4
Midwest	21	104	20.2	12	105	11.4
Southeast				318	2106	15.1

QI Measure 3: (Transcranial Doppler): Eligible SCD patients between ages of 2 -16, that had a Transcranial Doppler (TCD) screening within the last 15 months

Region	Q1			Q2		
	Numerator	Denominator	Percentage	Numerator	Denominator	Percentage
Heartland/Southwest	734	1786	41.1	758	1740	43.6
Midwest	299	371	80.6	389	477	81.6
Southeast				519	972	53.4

QI Measure 4a: (Immunization) Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[2]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8

1. Up to date; 2. Region where estimates may not be accurate due to confusion on how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9
Southeast[1]																					

1. Southeast did not provide vaccination data and is included in grey.
2. Detailed information to match flu season with quarter were provided during Q2. MOP is in process of being updated to reflect this.

QI Measure 4a Percentage Comparison Q1 vs Q2

Region	UTD[1] with PCV		UTD with PPSV		UTD with MenACYW		UTD with Hib		UTD with Flu		UTD with MenB		UTD with all	
	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %
Heartland/ Southwest	83.6	76.8	83.1	82.4	42.9	53.3	90.8	82.6	65.5	60.1	48.8	49.8	7.50	10.2
Midwest	69.1	76.8	78.2	86.3	77.4	78.0	68.3	67.1	67.3	62.8	89.5	66.8	43.8	38.9

Note: Southeast excluded since no vaccination data provided.

QI Measure 4b: (Immunization) Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Quarter 1 QI Measure 4b

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[2]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	151	295	51.5	20	263	7.60	13	263	4.94
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.16

1. Up to date; 2. Region where estimates may not be accurate due to confusion on how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 QI Measure 4b

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5
Southeast																					

1. Southeast did not provide vaccination data and is included in grey.
2. Detailed information to match flu season with quarter were provided during Q2. MOP is in process of being updated to reflect this.

QI Measure 4b Percentage Comparison Q1 vs Q2

Region	UTD[1] with PCV		UTD with PPSV		UTD with MenACYW		UTD with Hib		UTD with Flu		UTD with MenB		UTD with all	
	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %	Q1 %	Q2 %
Heartland/ Southwest	54.0	56.0	65.7	66.3	44.8	50.0	50.8	53.0	51.5	51.3	7.60	11.3	4.94	8.0
Midwest	55.0	57.8	68.3	73.5	73.5	76.0	50.8	55.4	60.8	58.8	69.3	68.1	30.7	26.5

Note: Southeast excluded since no vaccination data provided.

QI Measure 5: (Transitions to Adult Care) Number of patients that have a documented transition education

	Q1			Q2		
Region	**Numerator**	**Denominator**	**Percentage**	**Numerator**	**Denominator**	**Percentage**
Heartland/Southwest	81	165	49.1%	93	161	57.8
Midwest	9	48	18.8%	21	63	33.3
Southeast				132	329	40.1

QI Measure 6: (ECHO) Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Q1	Q2
Overall	109	161
Heartland/Southwest	61	43
Midwest	48	49
Southeast		69

Quarter 3 QI Data Summary

January 13, 2020

Overview: This report reflects data submitted for Quarter 3 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) which spanned July 1 to September 30, 2019. Regional Coordinating Centers (RCCs) aggregated and submitted data to CoLab between October 1 and November 15, 2019. Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers participation in ECHO

Regions collected data for this QI report from Electronic Health Records (EHR) or other medical records (such as manual chart review) at the site level. The sample for each region was intended to reflect the total number of sickle-cell patients seen within the specific quarter. For Quarter 3, four out of five regions submitted data (Heartland/Southwest, Midwest, Pacific and Southeast). The Northeast RCC is providing data on a 6-month schedule (reporting for Q2, Q4). Northeast RCC's data is included for Q1 (covers September 1, 2018 – February 28, 2019) and Q2 (covers January 1 to June 30, 2019) and therefore is not included in this report.

The first page of this report displays the aggregated values for the QI measures across regions. The following pages of this report provide QI measures and values by region. Notes about annotations are provided where relevant. Guidance to RCCs about inclusion for numerators and denominators is detailed in the Manual of Operating Procedures (MOP). Linked here is the most up-to-date MOP. Greyed out boxes indicate that a RCC did not provide data either because it was not applicable or collected.

Aggregated Values for Q3 QI Measures (*excluding QI Measure 4, immunizations- see below*)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[1]			QI Measure 5: Transitions to Adult Care[2]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	3654	5725	63.8	449	3961	11.3	2089	3798	55.0	297	778	38.2	201
Adult	1504	2424	62.0	463	2267	20.4							

Note: N = Numerator; D= Denominator

[1.] Only for eligible patients between 2 and 16 years of age
[2.] Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q3 QI Measure 4 (Immunization Status)[1]

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Pediatric	1358	1648	82.4	1349	1648	81.8	1111	1648	67.4	1291	1648	78.3	721	1648	43.8	1028	1639	62.7	337	1627	20.7
Adult	377	555	67.9	379	567	66.8	305	488	62.5	384	658	58.4	259	488	53.1	187	488	38.3	89	476	18.7

Note: N = Numerator; D= Denominator.

[1]Heartland/Southwest, Midwest, Pacific included in aggregate for immunizations.

*Vaccine priority for this project

Region Specific Values for QI Measures

QI Measure 1a: Hydroxyurea use among Children/Adolescents (Required measure for all RCCs to collect)

Percent of patients >9-months and <18 years of age prescribed Hydroxyurea – in the 6 months preceding September 30, 2019

Region	Q1			Q2			Q3		
	N	D	%	N	D	%	N	D	%
Heartland/Southwest	916	1959	46.8	942	1938	48.6	1035	1939	53.4
Midwest	345	437	78.9	338	393	86.0	418	537	77.8
Southeast				1552	2300	67.5	1913	2777	68.9
Pacific							288	468	61.5

QI Measure 1b: Hydroxyurea use among Adults (Required measure for all RCCs to collect)

Percent of patients 18 years and older prescribed Hydroxyurea – in the 6 months preceding September 30, 2019

Region	Q1			Q2			Q3		
	N	D	%	N	D	%	N	D	%
Heartland/Southwest	216	351	61.5	201	357	56.3	192	338	56.8
Midwest	75	108	69.4	90	119	75.6	96	119	80.7
Southeast				1157	1975	58.6	1117	1759	63.5
Pacific							99	208	47.6

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents (Optional Measure)

Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU– in the 6 months preceding September 30, 2019

Region	Q1			Q2			Q3		
	N	D	%	N	D	%	N	D	%
Heartland/Southwest	124	1220	10.2	120	1085	11.1	126	1010	12.5
Midwest	54	413	13.1	58	540	10.7	58	482	12.0
Southeast				204	1402	14.6	253	2402	10.5
Pacific							12	67	17.9

QI Measure 2b: Disease modifying therapy use other than HU among Adults (Optional Measure)

Percent of patients 18 years and older prescribed disease modifying therapy other than HU – in the 6 months preceding September 30, 2019

Region	Q1			Q2			Q3		
	N	D	%	N	D	%	N	D	%
Heartland/Southwest	80	250	32.0	91	237	38.4	96	237	40.5
Midwest	21	104	20.2	12	105	11.4	13	105	12.4
Southeast				318	2106	15.1	350	1909	18.3
Pacific							4	16	25.0

QI Measure 3: Transcranial Doppler (Optional Measure)

Eligible SCD patients between ages of 2-16, that had a Transcranial Doppler (TCD) screening within the 15 months prior to the end of the quarter

Region	Q1			Q2			Q3		
	N	D	%	N	D	%	N	D	%
Heartland/Southwest	734	1786	41.1	758	1740	43.6	934	2099	44.5
Midwest	299	371	80.6	389	477	81.6	393	472	83.3
Southeast				519	972	53.4	540	920	58.7
Pacific							222	307	72.3

QI Measure 4a: Immunization

Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[3]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8

1. UTD: Up to date. This footnote will not be repeated in future tables.
2. Detailed information to match flu season with quarter were provided during Q2. This footnote will not be repeated in future tables.
3. Region where estimates may not be accurate due to need to clarify how to report data if not all sites reported immunization data. Issue has since been resolved.

Quarter 2 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9

Quarter 3 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	812	997	81.4	843	997	84.6	596	997	59.8	858	997	86.1	347	997	34.8	531	997	53.3	121	997	12.1
Midwest	534	630	84.8	494	630	78.4	501	630	79.5	428	630	67.9	361	630	57.3	487	630	77.3	216	630	34.3
Pacific	12	21	57.1	12	21	57.1	14	21	66.7	5	21	23.8	13	21	61.9	10	12	83.3			

QI Measure 4a Percentage Comparison Q2 vs Q3

Region	UTD with PCV		UTD with PPSV		UTD with MenACYW		UTD with Hib		UTD with Flu		UTD with MenB		UTD with all	
	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %
Heartland/ Southwest	76.8	81.4	82.4	84.6	53.3	59.8	82.6	86.1	60.1	34.8	49.8	53.3	10.2	12.1
Midwest	76.8	84.8	86.3	78.4	78.0	79.5	67.1	67.9	62.8	57.3	66.8	77.3	38.9	34.3

Note: Southeast excluded since no vaccination data provided (in either quarter). Pacific excluded since Q2 data is pending. Northeast on 6-month schedule for data collection and therefore not included for Q3 data.

QI Measure 4b: (Immunization) Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Quarter 1 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[1]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	151	295	51.5	20	263	7.60	13	263	4.94
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.16

1. Region where estimates may not be accurate due to need to clarify how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5

Quarter 3 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	184	264	69.7	200	264	75.8	153	264	58.0	159	264	60.2	122	264	46.2	59	264	22.3	31	264	11.7
Midwest	170	212	80.2	140	212	66.0	148	212	69.8	100	212	47.2	131	212	61.8	125	212	59.0	58	212	27.4
Pacific	23	79	29.1	39	91	42.9	4	12	33.3	125	182	68.7	6	12	50.0	3	12	25.0			

QI Measure 4b Percentage Comparison Q2 vs Q3

Region	UTD with PCV		UTD with PPSV		UTD with MenACYW		UTD with Hib		UTD with Flu		UTD with MenB		UTD with all	
	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %	Q2 %	Q3 %
Heartland/ Southwest	56.0	69.7	66.3	75.8	50.0	58.0	53.0	60.2	51.3	46.2	11.3	22.3	8.0	11.7
Midwest	57.8	80.2	73.5	66.0	76.0	69.8	55.4	47.2	58.8	61.8	68.1	59.0	26.5	27.4

Note: Southeast excluded since no vaccination data provided (in either quarters). Pacific excluded since Q2 data is pending. Northeast on a 6-month schedule and not providing data for Q3, therefore is not in this comparison chart.

QI Measure 5: (Transitions to Adult Care) Number of patients that have a documented transition education

	Q1			Q2			Q3		
Region	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Heartland/Southwest	81	165	49.1	93	161	57.8	117	184	63.6
Midwest	9	48	18.8	21	63	33.3	16	63	25.4
Pacific							13	19	68.4
Southeast				132	329	40.1	151	512	29.5

QI Measure 6: ECHO (Count of providers participating in project Echo meetings or Tele-mentoring calls)

Region	**Q1**	**Q2**	**Q3**
Overall	109	161	149
Heartland/Southwest	61	43	58
Midwest	48	49	35
Pacific			52
Southeast		69	56

Quarter 4 Quality Improvement Data Summary

April 1, 2020

Table of Contents

Overview: This report reflects data submitted for Quarter 4 2019 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) which spanned October 1 to December 31, 2019. Regional Coordinating Centers (RCCs) aggregated and submitted data to CoLab between January 1 and February 15, 2020.

Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers' participation in ECHO

Regions collected data for this QI report via reporting from Electronic Health Records (EHR) and/or by manual chart review at the site level. For Quarter 4, all five regions submitted data (Heartland/Southwest, Midwest, Northeast, Pacific and Southeast). The Northeast RCC is providing data on a 6-month schedule (reporting for Q2, Q4). *

The first page of this report displays the aggregated values for the QI measures across regions by quarter and biannually based on how frequently data was submitted. The following pages provide QI measures and values by region.

The sample for each region was intended to reflect the total number of sickle-cell patients seen within the specific quarter. All data, except for immunizations, reflect a population-level view. Greyed out boxes indicate that a RCC did not provide data either because it was not applicable or not collected. Notes about annotations are provided where relevant. Guidance to RCCs about inclusion for numerators and denominators is detailed in the Manual of Operating Procedures (MOP). Linked here is the most up-to- date MOP.

*The Northeast RCC is providing data on a 6-month schedule (reporting for Q2, Q4 only). Therefore, data for all regions are shown for Q1+2 together and then Q3+4 together.

Aggregated Values for Q4 QI Measures (excluding QI Measure 4, immunizations- see below)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[1]			QI Measure 5: Transitions to Adult Care[2]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	3479	4846	71.7	552	3874	14.2	1966	2751	71.5	372	941	39.5	220
Adult	1590	2603	61.1	551	2466	22.3							

Note: N = Numerator; D= Denominator

1. Only for eligible patients between 2 and 16 years of age
2. Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q4 QI Measure 4 (Immunization Status)

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	1443	1653	87.3	1387	1648	84.2	1178	1644	71.7	1365	1653	82.6	943	1653	57.0	1092	1415	77.2	487	1404	34.7
Adult	339	498	68.1	375	514	73.0	292	432	67.6	382	600	63.7	192	432	44.4	179	400	44.8	65	384	16.9

Note: N = Numerator; D= Denominator.

*Vaccine priority for this project

Only Heartland/Southwest, Midwest, Pacific included in aggregate for Q4.

Aggregation Across Quarters: Q1 – Q2 (January 1-June 30, 2019)[1]

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	5430	9118	59.6	635	5063	12.5	3423	6519	52.5	434	986	44.0	568
Adult	2413	4225	57.1	685	3403	20.1							

	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3082	3970	77.6	3111	3770	82.5	2406	3977	60.5	3125	3989	78.3	2202	3954	55.7	1819	3664	49.6	797	3693	21.6
Adult	1189	2066	57.6	1271	2129	59.7	1103	1952	56.5	1287	2304	55.9	743	1950	38.1	371	1918	19.3	419	1889	22.2

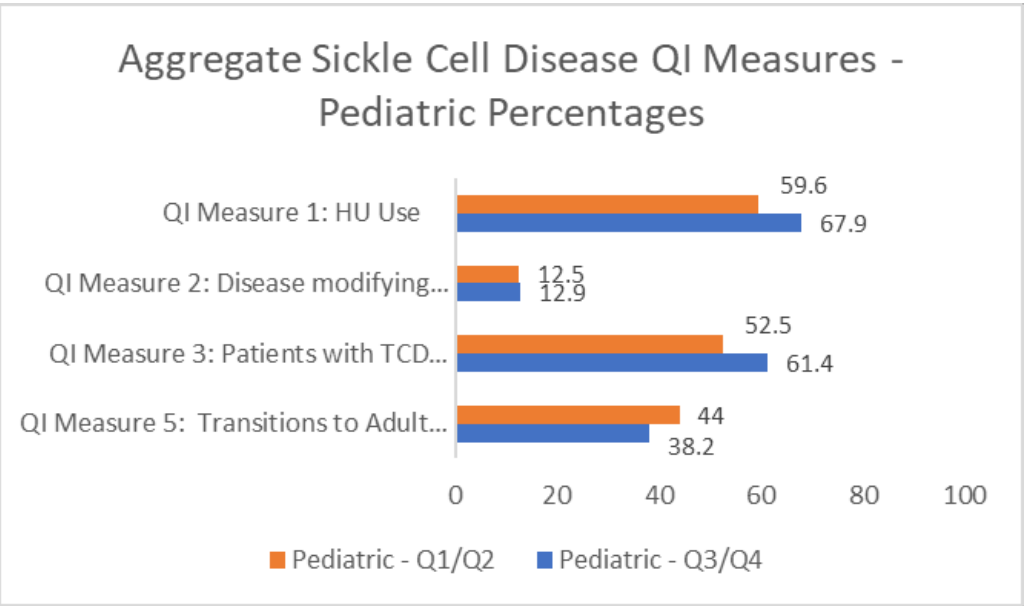
Aggregation Across Quarters: Q3 – Q4 (July 1-December 31, 2019)[1]

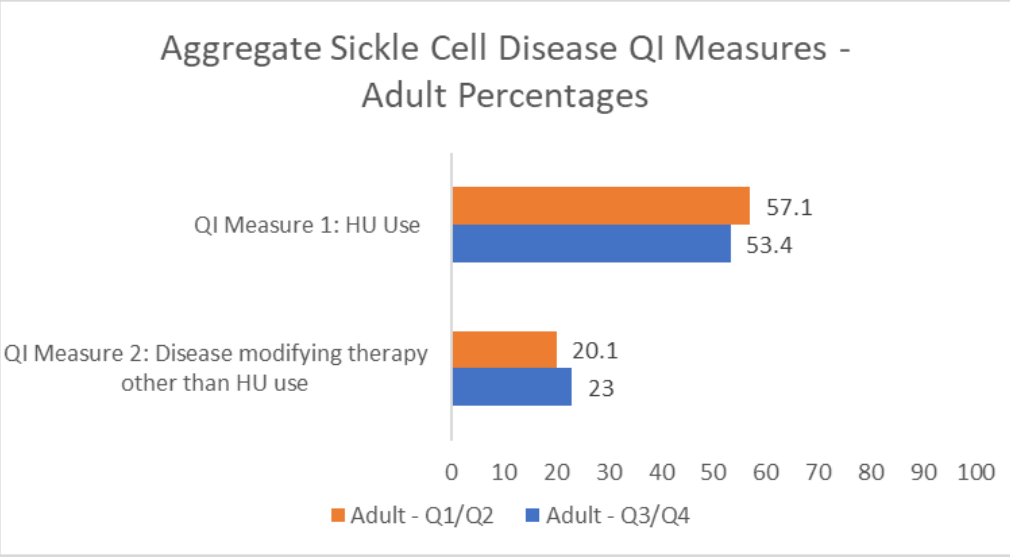
	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	7553	11121	67.9	1029	7989	12.9	4355	7090	61.4	710	1860	38.2	567
Adult	3669	5964	61.5	1185	5144	23.0							

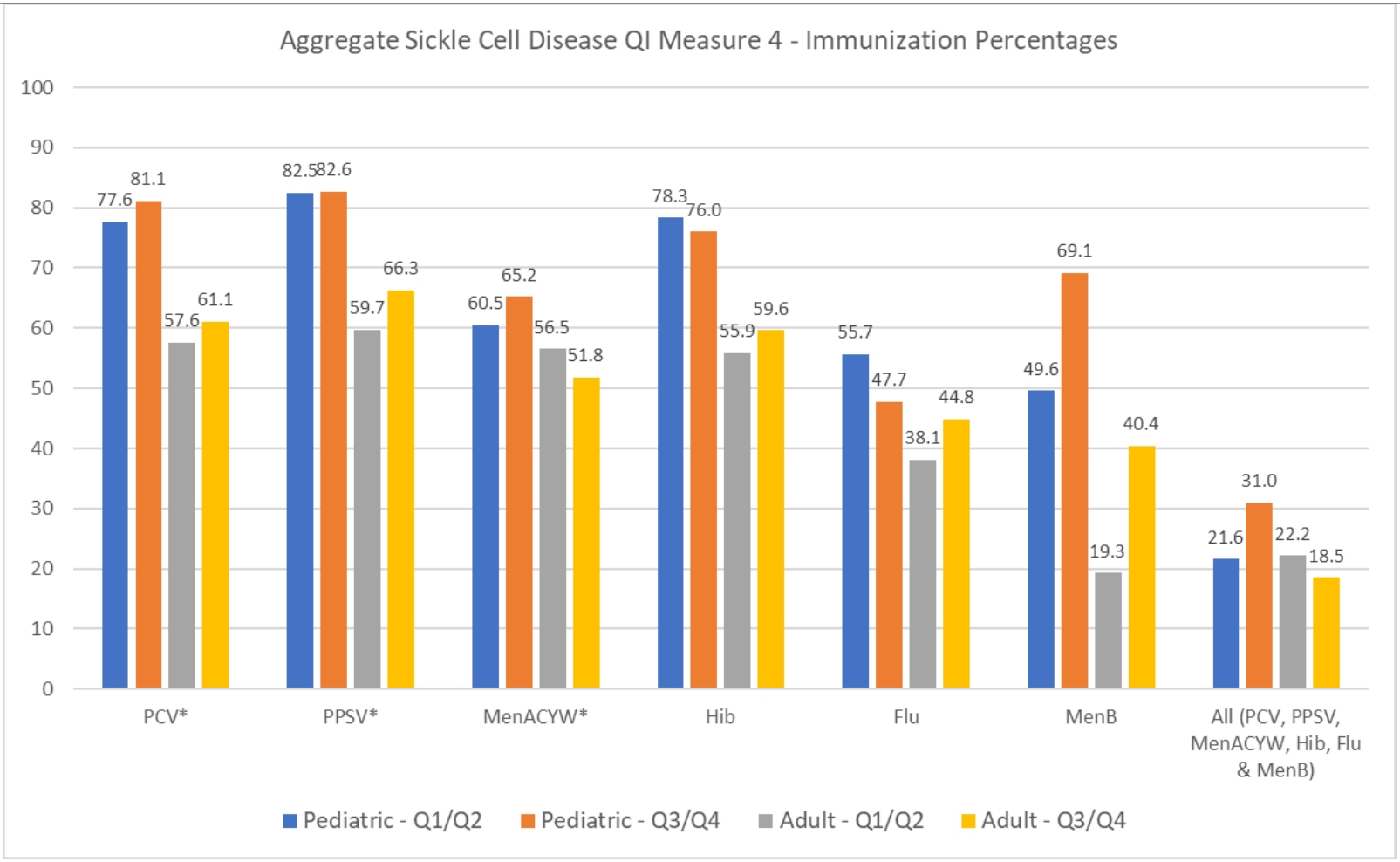
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3355	4135	81.1	3407	4123	82.6	2688	4122	65.2	3119	4103	76.0	1971	4135	47.7	2291	3317	69.1	1031	3322	31.0
Adult	954	1561	61.1	1074	1619	66.3	739	1428	51.8	821	1378	59.6	640	1428	44.8	407	1008	40.4	181	980	18.5

1. See introduction text on page 1 for more detail on aggregation across quarters

Graphs of Aggregation Across Quarters






Aggregate Sickle Cell Disease QI Measure 4 - Immunization Percentages
100
90
80
70
60
50
40
30
20
10
0
77.6
81.1
57.6
61.1
82.5
82.6
59.7
66.3
60.5
65.2
56.5
51.8
78.3
76.0
55.9
59.6
55.7
47.7
38.1
44.8
49.6
69.1
19.3
40.4
21.6
31.0
22.2
18.5
PCV*
PPSV*
MenACYW*
Hib
Flu
MenB
All (PCV, PPSV, MenACYW, Hib, Flu & MenB)
Pediatric - Q1/Q2
Pediatric - Q3/Q4
Adult - Q1/Q2
Adult - Q3/Q4

Region Specific Values for QI Measures

QI Measure 1a: Hydroxyurea use among Children/Adolescents (Required measure for all RCCs to collect)

Percent of patients >9-months and <18 years of age prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019		
	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	916	1959	46.8	942	1938	48.6	1035	1939	53.4	863	1156	74.7
Midwest	345	437	78.9	338	393	86.0	418	537	77.8	361	439	82.2
Northeast[1]	643	883	72.8	775	1179	65.7				429	597	71.9
Southeast				1552	2300	67.5	1910	2734	69.9	1958	2735	71.6
Pacific	277	453	61.2	285	459	62.1	288	468	61.5	291	516	56.4

1. Note, Northeast Q1 data included September 1, 2018 – February 28th, 2019. In order to compare across all RCCs, NE data for Q2 (Combined Q1/Q2: January 1, 2019 – June 30, 2019) and Q4 (Combined Q3/Q4: July 1, 2019 – December 31, 2019) were used as proxy for the data presented for across quarters. This footnote will not be repeated for each measure, however, is applicable across all.

QI Measure 1b: Hydroxyurea use among Adults (Required measure for all RCCs to collect)

Percent of patients 18 years and older prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019		
	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	216	351	61.5	201	357	56.3	192	338	56.8	229	380	60.3
Midwest	75	108	69.4	90	119	75.6	96	119	80.7	93	119	78.2
Northeast	485	874	55.5	437	891	48.7				541	870	62.2
Southeast				1157	1975	58.6	1151	1826	63.0	1156	1876	61.6
Pacific	128	216	59.3	109	208	52.4	99	208	47.6	112	228	49.1

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents (Optional Measure)

Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019		
	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	124	1220	10.2	120	1085	11.1	126	1010	12.5	104	389	26.7
Midwest	54	413	13.1	58	540	10.7	58	482	12.0	86	539	16.0
Northeast	62	275	22.5	51	273	18.7				26	197	13.2
Southeast				204	1402	14.6	255	2359	10.8	350	2864	12.2
Pacific	12	65	18.5	12	65	18.5	12	67	17.9	12	82	14.6

QI Measure 2b: Disease modifying therapy use other than HU among Adults (Optional Measure)

Percent of patients 18 years and older prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019		
	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	80	250	32.0	91	237	38.4	96	237	40.5	105	254	41.3
Midwest	21	104	20.2	12	105	11.4	13	105	12.4	20	103	19.4
Northeast	161	406	39.7	154	576	26.7				165	497	33.2
Southeast				318	2106	15.1	356	1823	19.5	424	2099	20.2
Pacific	4	12	33.3	5	13	38.5	4	16	25.0	2	10	20.0

QI Measure 3: Transcranial Doppler (Optional Measure)

Eligible SCD patients between ages of 2-16, that had a Transcranial Doppler (TCD) screening within the 15 months prior to the end of the quarter

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019		
	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	734	1786	41.1	758	1740	43.6	934	2099	44.5	787	1030	76.4
Midwest	299	371	80.6	389	477	81.6	393	472	83.3	415	523	79.3
Northeast	393	881	44.6	295	584	50.5				300	541	55.5
Southeast				519	972	53.4	540	920	58.7	540	861	62.7
Pacific	214	283	75.6	215	306	70.3	222	307	72.3	224	337	66.5

QI Measure 4a: Immunization - Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[3]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8
Northeast	598	1493	40.1	754	1493	50.5	448	1493	30.0	525	1493	35.2	316	1493	21.2	38	1493	2.5	119	1493	8.0
Southeast																					
Pacific	18	29	62.1	17	29	58.6	20	29	69.0	6	29	20.7	16	29	55.2	13	29	44.8			

1. UTD= Up to date. This footnote will not be repeated in future tables.
2. Detailed information to match flu season with quarter were provided during Q2. This footnote will not be repeated in future tables.
3. Region where estimates may not be accurate due to need to clarify how to report data if not all sites reported immunization data. Issue has since been resolved.

Quarter 2 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9
Northeast	815	1024	79.6	854	1024	83.4	620	1024	60.5	818	1024	79.9	344	1024	33.6	197	1024	19.2	182	1024	17.8
Southeast																					
Pacific	14	21	66.7	11	21	52.4	15	21	71.4	3	21	14.3	17	21	81.0	10	21	47.6			

Quarter 3 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	812	997	81.4	843	997	84.6	596	997	59.8	858	997	86.1	347	997	34.8	531	997	53.3	121	997	12.1
Midwest	534	630	84.8	494	630	78.4	501	630	79.5	428	630	67.9	361	630	57.3	487	630	77.3	216	630	34.3
Northeast																					
Southeast																					
Pacific	12	21	57.1	12	21	57.1	14	21	66.7	5	21	23.8	13	21	61.9	10	12	83.3			

Quarter 4 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	824	968	85.1	842	968	87.0	638	968	65.9	855	968	88.3	472	968	48.8	495	743	66.6	190	743	25.6
Midwest	600	661	90.8	528	656	80.5	523	652	80.2	496	661	75.0	456	661	69.0	580	648	89.5	297	661	44.9
Northeast	554	834	66.4	617	827	81.1	399	830	48.1	463	802	57.7	307	834	36.8	171	263	65.0	207	291	71.1
Southeast																					
Pacific	19	24	79.2	17	24	70.8	17	24	70.8	14	24	58.3	15	24	62.5	17	24	70.8			

QI Measure 4b: Immunization - Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series
Quarter 1 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[1]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	152	295	51.5	20	263	7.60	13	263	4.94
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.2
Southeast																					
Pacific	11	94	11.7	35	111	31.5	10	17	58.8	142	191	74.3	10	17	58.8	7	17	41.2			

1. Region where estimates may not be accurate due to need to clarify how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5
Northeast	625	933	67.0	525	933	56.3	511	933	54.8	485	933	52.0	186	933	19.9	36	933	3.9	270	933	28.9
Southeast																					
Pacific	21	83	25.3	38	95	40.0	5	12	41.7	141	190	74.2	6	12	50.0	4	12	33.3			

Quarter 3 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	184	264	69.7	200	264	75.8	153	264	58.0	159	264	60.2	122	264	46.2	59	264	22.3	31	264	11.7
Midwest	170	212	80.2	140	212	66.0	148	212	69.8	100	212	47.2	131	212	61.8	125	212	59.0	58	212	27.4
Northeast																					
Southeast																					
Pacific	23	79	29.1	39	91	42.9	4	12	33.3	125	182	68.7	6	12	50.0	3	12	25.0			

Quarter 4 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	134	203	66.0	179	203	88.2	132	203	65.0	128	203	63.1	94	203	46.3	36	171	21.1	20	171	11.7
Midwest	179	213	84.0	151	213	70.9	157	213	73.7	110	213	51.6	91	213	42.7	137	213	64.3	45	213	21.1
Northeast	238	508	46.9	320	538	59.5	142	508	28.0	55	120	45.8	189	508	37.2	41	120	34.2	27	120	22.5
Southeast																					
Pacific	26	82	31.7	45	98	45.9	3	16	18.8	144	184	78.3	7	16	43.8	6	16	37.5			

QI Measure 5: Transitions to Adult Care - Number of patients that have a documented transition education

	Q1 2019			Q2 2019			Q3 2019			Q4 2019		
Region	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Heartland/Southwest	81	165	49.1	93	161	57.8	117	184	63.6	118	181	65.2
Midwest	9	48	18.8	21	63	33.3	16	63	25.4	18	59	30.5
Northeast	21	110	19.1	68	175	38.9				6	91	6.6
Pacific	16	23	69.6	14	22	63.6	13	19	68.4	11	18	61.1
Southeast				132	329	40.1	186	562	33.1	225	683	32.9

QI Measure 6: ECHO - Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Q1 2019	Q2 2019	Q3 2019	Q4 2019	Total 2019
Heartland/Southwest	61	43	58	54	216
Midwest	48	49	35	46	178
Northeast	170	242		146	558
Pacific	59		52	65	176
Southeast		66	56	55	177

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Quarter 1 2020 Quality Improvement Data Summary

June 25, 2020

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Overview: This report reflects data submitted for Quarter 1 2020 QI data collection for the SCDTDRCP which spanned January 1 to March 31, 2020. Regional Coordinating Centers (RCCs) aggregated and submitted data to CoLab between April 1 and May 15, 2020.

Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers' participation in ECHO

Regions collected data for this QI report via reporting from Electronic Health Records (EHR) and/or by manual chart review at the site level. For Quarter 1 2020, four regions submitted data (Heartland/Southwest, Midwest, Pacific and Southeast). The Northeast RCC is providing data on a 6-month schedule (reporting for Q2, Q4).

The first page of this report displays the aggregated values for the QI measures across regions by quarter and biannually based on how frequently data was submitted. The following pages provide QI measures and values by region.

The sample for each region was intended to reflect the total number of sickle-cell patients seen within the specific quarter. All data, except for immunizations, reflect a population-level view. Greyed out boxes indicate that an RCC did not provide data either because it was not applicable or not collected. Notes about annotations are provided where relevant. Headings for Q1 2020 data are highlighted in green to assist in readability across quarters. Guidance to RCCs about inclusion for numerators and denominators is detailed in the Manual of Operating Procedures (MOP). Linked here is the most up-to- date MOP.

Acknowledgement of the impact of Covid-19: Starting in March 2020, the COVID-19 Pandemic reached the United States, strongly impacting multiple regions. Many of the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) providers at the local and regional levels were engaged in either front-line care or planning and developing new procedures and processes to respond to novel needs. All SCDTDRCP regions reported upheaval in their clinical systems and concern that appointments, both elective and/or essential, were not happening for SCD patients as usual or recommended. As such, there is some known (limited sites have not been able to report due to staff reductions) and potentially unknown (reduced or eliminated visits, clinical priority shifts) variation in this quarter's data. We anticipate variation may continue into future reports.

Aggregated Values for Q1 2020 QI Measures (excluding QI Measure 4, immunizations- see below)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[1]			QI Measure 5: Transitions to Adult Care[2]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	3518	5012	70.2	417	3348	12.5	1899	2743	69.2	366	946	38.7	315
Adult	1277	2416	52.9	466	2210	21.1							

Note: N = Numerator; D= Denominator

[1.] Only for eligible patients between 2 and 16 years of age
[2.] Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q1 2020 QI Measure 4 (Immunization Status)[1]

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	1547	1773	87.3	1358	1626	83.5	1136	1649	68.9	1415	1717	82.4	1123	1780	63.1	783	1114	70.3	641	1548	41.4
Adult	404	606	66.7	419	623	67.3	318	541	58.8	435	712	61.1	253	541	46.8	211	502	42.0	88	485	18.1

Note: N = Numerator; D= Denominator.

1. Only Heartland/Southwest, Midwest, Pacific included in aggregate for Q1 2020 Measure 4 Immunization Status. The SE region does not collect immunizations (adult or pediatric).

*Vaccine priority for this project

Region Specific Values for QI Measures

QI Measure 1a: Hydroxyurea use among Children/Adolescents (Required measure for all RCCs to collect)

Percent of patients >9-months and <18 years of age prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020		
	N	D	%	N	N	D	%	D	%	N	D	%	N	D	%
Heartland/Southwest	916	1959	46.8	942	1938	48.6	1035	1939	53.4	863	1156	74.7	927	1258	73.7
Midwest	345	437	78.9	338	393	86.0	418	537	77.8	361	439	82.2	413	508	81.3
Northeast[1]	643	883	72.8	775	1179	65.7				429	597	71.9			
Southeast				1552	2300	67.5	1910	2734	69.9	1958	2735	71.6	1972	2877	68.5
Pacific	277	453	61.2	285	459	62.1	288	468	61.5	291	516	56.4	206	369	55.8

1. Note, Northeast Q1 data included September 1, 2018 – February 28th, 2019. In order to compare across all RCCs, NE data for Q2 (Combined Q1/Q2: January 1, 2019 – June 30, 2019) and Q4 (Combined Q3/Q4: July 1, 2019 – December 31, 2019) were used as proxy for the data presented for across quarters. This footnote will not be repeated for each measure, however, is applicable across all.

QI Measure 1b: Hydroxyurea use among Adults (Required measure for all RCCs to collect)

Percent of patients 18 years and older prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	216	351	61.5	201	357	56.3	192	338	56.8	229	380	60.3	256	469	54.6
Midwest	75	108	69.4	90	119	75.6	96	119	80.7	93	119	78.2	84	111	75.7
Northeast	485	874	55.5	437	891	48.7				541	870	62.2			
Southeast				1157	1975	58.6	1151	1826	63.0	1156	1876	61.6	828	1605	51.6
Pacific	128	216	59.3	109	208	52.4	99	208	47.6	112	228	49.1	109	231	47.2

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents (Optional Measure)

Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	124	1220	10.2	120	1085	11.1	126	1010	12.5	104	389	26.7	91	408	22.3
Midwest	54	413	13.1	58	540	10.7	58	482	12.0	86	539	16.0	23	198	11.6
Northeast	62	275	22.5	51	273	18.7				26	197	13.2			
Southeast				204	1402	14.6	255	2359	10.8	350	2864	12.2	303	2742	11.1
Pacific	12	65	18.5	12	65	18.5	12	67	17.9	12	82	14.6			

QI Measure 2b: Disease modifying therapy use other than HU among Adults (Optional Measure)

Percent of patients 18 years and older prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	80	250	32.0	91	237	38.4	96	237	40.5	105	254	41.3	123	342	36.0
Midwest	21	104	20.2	12	105	11.4	13	105	12.4	20	103	19.4	18	83	21.7
Northeast	161	406	39.7	154	576	26.7				165	497	33.2			
Southeast				318	2106	15.1	356	1823	19.5	424	2099	20.2	325	1785	18.2
Pacific	4	12	33.3	5	13	38.5	4	16	25.0	2	10	20.0			

QI Measure 3: Transcranial Doppler (Optional Measure)

Eligible SCD patients between ages of 2-16, that had a Transcranial Doppler (TCD) screening within the 15 months prior to the end of the quarter

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	734	1786	41.1	758	1740	43.6	934	2099	44.5	787	1030	76.4	773	1144	67.6
Midwest	299	371	80.6	389	477	81.6	393	472	83.3	415	523	79.3	358	442	81.0
Northeast	393	881	44.6	295	584	50.5				300	541	55.5			
Southeast				519	972	53.4	540	920	58.7	540	861	62.7	561	832	67.4
Pacific	214	283	75.6	215	306	70.3	222	307	72.3	224	337	66.5	207	325	63.7

QI Measure 4a: Immunization - Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[3]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8
Northeast	598	1493	40.1	754	1493	50.5	448	1493	30.0	525	1493	35.2	316	1493	21.2	38	1493	2.5	119	1493	8.0
Southeast																					
Pacific	18	29	62.1	17	29	58.6	20	29	69.0	6	29	20.7	16	29	55.2	13	29	44.8			

1. UTD= Up to date. This footnote will not be repeated in future tables.
2. Detailed information to match flu season with quarter were provided during Q2 2019. This footnote will not be repeated in future tables.
3. Region where estimates may not be accurate due to need to clarify how to report data if not all sites reported immunization data. Issue has since been resolved.

Quarter 2 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9
Northeast	815	1024	79.6	854	1024	83.4	620	1024	60.5	818	1024	79.9	344	1024	33.6	197	1024	19.2	182	1024	17.8
Southeast																					
Pacific	14	21	66.7	11	21	52.4	15	21	71.4	3	21	14.3	17	21	81.0	10	21	47.6			

Quarter 3 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	812	997	81.4	843	997	84.6	596	997	59.8	858	997	86.1	347	997	34.8	531	997	53.3	121	997	12.1
Midwest	534	630	84.8	494	630	78.4	501	630	79.5	428	630	67.9	361	630	57.3	487	630	77.3	216	630	34.3
Northeast																					
Southeast																					
Pacific	12	21	57.1	12	21	57.1	14	21	66.7	5	21	23.8	13	21	61.9	10	12	83.3			

Quarter 4 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	824	968	85.1	842	968	87.0	638	968	65.9	855	968	88.3	472	968	48.8	495	743	66.6	190	743	25.6
Midwest	600	661	90.8	528	656	80.5	523	652	80.2	496	661	75.0	456	661	69.0	580	648	89.5	297	661	44.9
Northeast	554	834	66.4	617	827	81.1	399	830	48.1	463	802	57.7	307	834	36.8	171	263	65.0	207	291	71.1
Southeast																					
Pacific	19	24	79.2	17	24	70.8	17	24	70.8	14	24	58.3	15	24	62.5	17	24	70.8			

Quarter 1 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	934	1102	84.8	944	1102	85.7	722	1102	65.5	970	1102	88.0	646	1102	58.6	618	876	70.5	341	876	38.9
Midwest	594	650	91.4	397	503	78.9	396	526	75.3	430	594	72.4	461	657	70.2	147	217	67.7	300	672	44.6
Northeast																					
Southeast																					
Pacific	19	21	90.5	17	21	81.0	18	21	85.7	15	21	71.4	16	21	76.2	18	21	85.7			

QI Measure 4b: Immunization - Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[1]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	152	295	51.5	20	263	7.60	13	263	4.94
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.2
Southeast																					
Pacific	11	94	11.7	35	111	31.5	10	17	58.8	142	191	74.3	10	17	58.8	7	17	41.2			

1. Region where estimates may not be accurate due to need to clarify how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5
Northeast	625	933	67.0	525	933	56.3	511	933	54.8	485	933	52.0	186	933	19.9	36	933	3.9	270	933	28.9
Southeast																					
Pacific	21	83	25.3	38	95	40.0	5	12	41.7	141	190	74.2	6	12	50.0	4	12	33.3			

Quarter 3 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	184	264	69.7	200	264	75.8	153	264	58.0	159	264	60.2	122	264	46.2	59	264	22.3	31	264	11.7
Midwest	170	212	80.2	140	212	66.0	148	212	69.8	100	212	47.2	131	212	61.8	125	212	59.0	58	212	27.4
Northeast																					
Southeast																					
Pacific	23	79	29.1	39	91	42.9	4	12	33.3	125	182	68.7	6	12	50.0	3	12	25.0			

Quarter 4 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	134	203	66.0	179	203	88.2	132	203	65.0	128	203	63.1	94	203	46.3	36	171	21.1	20	171	11.7
Midwest	179	213	84.0	151	213	70.9	157	213	73.7	110	213	51.6	91	213	42.7	137	213	64.3	45	213	21.1
Northeast	238	508	46.9	320	538	59.5	142	508	28.0	55	120	45.8	189	508	37.2	41	120	34.2	27	120	22.5
Southeast																					
Pacific	26	82	31.7	45	98	45.9	3	16	18.8	144	184	78.3	7	16	43.8	6	16	37.5			

Quarter 1 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	201	310	64.8	219	310	70.6	156	310	50.3	176	310	56.8	151	310	48.7	68	271	25.1	44	271	16.2
Midwest	176	214	82.2	150	214	70.1	156	214	72.9	110	214	51.4	93	214	43.5	138	214	64.5	44	214	20.6
Northeast																					
Southeast																					
Pacific	27	82	32.9	50	99	50.5	6	17	35.3	149	188	79.3	9	17	52.9	5	17	29.4			

QI Measure 5: Transitions to Adult Care - Number of patients that have a documented transition education

	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020		
Region	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Heartland/Southwest	81	165	49.1	93	161	57.8	117	184	63.6	118	181	65.2	108	173	62.4
Midwest	9	48	18.8	21	63	33.3	16	63	25.4	18	59	30.5	20	70	28.8
Northeast	21	110	19.1	68	175	38.9				6	91	6.6			
Pacific	16	23	69.6	14	22	63.6	13	19	68.4	11	18	61.1	12	17	70.6
Southeast				132	329	40.1	186	562	33.1	225	683	32.9	226	686	32.9

QI Measure 6: ECHO - Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Q1 2019	Q2 2019	Q3 2019	Q4 2019	Q1 2020
Heartland/Southwest	61	43	58	54	35
Midwest	48	49	35	46	120
Northeast	170	242		146	
Pacific	59		52	65	88
Southeast		66	56	55	72

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Quality Improvement Data Summary

Quarter 2 2020

September 25, 2020

Prepared by:

NICHQ's Department of Applied Research and Evaluation (DARE)

Executive Summary

To address the ongoing challenges of serving children and adults with sickle cell disease (SCD), and to improve care and outcomes, Congress created legislation that has funded the SCDTDRCP since 2003. The goals of the SCDTDRCP are to 1) improve coordination and service delivery for individuals living with SCD; 2) improve access to services; and 3) improve and expand on provider knowledge of SCD treatment and care. The SCDTDRCP is organized into five Regional Coordinating Centers (RCCs) and one National Coordinating Center (NCC).

RCCs are charged with quarterly reporting on at least two Quality Improvement (QI) Measures. QI data is collected on a quarterly basis. All regions must assess Hydroxyurea (HU) use; the regions can choose which other QI measure(s) to collect. The six QI measures include: 1) Hydroxyurea use; 2) Other disease modifying therapy use; 3) Immunization status; 4) Transcranial Doppler Ultrasound screening; 5) Transitions to adult care; and 6) Providers' participation in Extension of Community Healthcare Outcomes (ECHO).

Data in this report includes aggregate data from all regions reporting for the quarter, aggregate data across multiple quarters to provide population level view, and regional data by quarter.

As of Quarter 2 (June 1-August 31) 2020, the RCCs have been collecting data for six quarters. Some observations thus far include:

- RCCs are centers of excellence that have made strides in implementation of guideline-based care for SCD.
- Prescribing of HU and TCD screening remain measures that RCCs are capturing consistently.
- ECHO has become a standard practice for all regions, and attendance for these sessions have been consistently tracked.
- Capturing immunization has been a challenge for the majority of clinical sites due to inconsistencies among how this data is captured via state records, EMR/chart documentation and/or patient recall.
- The definition of transition plan to adult care is not always consistent among sites. As well, consistent documentation of a transition meeting(s) varies, leading to concerns of under reporting. However, sites that have been able to define transition planning and have an infrastructure to capture this component have had success in collecting this QI measure.

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Overview: This report reflects data submitted for Quarter 2 2020 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) which spanned April 1 to June 30, 2020. Regional Coordinating Centers (RCCs) aggregated and submitted data to CoLab between July 1 and August 15, 2020.

Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers' participation in ECHO

Regions collected data for this QI report via reporting from Electronic Health Records (EHR) and/or by manual chart review at the site level. For Quarter 2 2020, five regions submitted data (Heartland/Southwest, Midwest, Northeast, Pacific and Southeast). Four of the five RCCs collect data on a quarterly basis. The Northeast RCC provides data on a 6-month schedule (reporting for Q2, Q4). All data is re-run on a 6-month basis (at Q2 and Q4) and any updates made to previous quarterly data will be reflected at these intervals. The last re-run of all data occurred on **8/19/20**.

The first page of this report displays the aggregated values for the QI measures across regions for Q2 2020 and then presents biannual aggregate values (i.e., combining Quarters 1 & 2 and Quarters 3 & 4) based on how frequently regional data was submitted. Starting on page eight, these biannual aggregate values across quarters are included to provide a population-level perspective of QI measures. These are also displayed in graphs. Pages 12 through 20 provide QI measures by region.

The sample for each region reflects the total number of sickle-cell patients seen from participating sites of a region within the specific quarter. Greyed out boxes indicate that an RCC did not provide data either because it was not applicable or not collected. Notes about annotations are provided where relevant. Headings for the current Q2 2020 data are highlighted in green to assist in readability across quarters. Guidance to RCCs about inclusion for numerators and denominators is detailed in the Manual of Operating Procedures (MOP). Linked here is the most up-to-date MOP.

Acknowledgement of the impact of COVID-19: Starting in March 2020, the COVID-19 Pandemic reached the United States. COVID-19 has impacted all regions. Many of the SCDTDRCP providers at the local and regional levels were engaged in either front-line care or planning and developing new procedures and processes to respond to emerging needs. All SCDTDRCP regions reported upheaval in their clinical systems and concern that appointments, both elective and/or essential, were not occurring for SCD patients as usual or as recommended. Therefore, there is some known (i.e., certain sites have been unable to report due to staff reductions) and potentially unknown (i.e., reduced or eliminated visits, decisions not to change medication during this time, clinical priority shifts) variation in all 2020 data. We anticipate continued variation in future reports as the COVID-19 pandemic continues.

Aggregated Values for Q2 2020 QI Measures (excluding QI Measure 4, immunizations- see below)[1]

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[2]			QI Measure 5: Transitions to Adult Care[3]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	3644	5127	71.1	476	3577	13.3	2065	3056	67.6	376	908	41.0	639
Adult	1537	2717	56.6	513	2490	20.6							

Note: N = Numerator; D= Denominator

1. Data included in aggregate values for Q2 2020 includes four regions (Heartland/Southwest, Midwest, Pacific and Southeast).
2. Only for eligible patients between 2 and 16 years of age
3. Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q2 2020 QI Measure 4 (Immunization Status)[1]

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	1513	1715	88.2	1330	1559	85.3	1188	1608	73.9	1375	1658	83.0	1070	1701	62.9	844	1074	78.6	615	1487	41.4
Adult	442	643	68.7	437	656	66.6	338	569	59.4	456	733	62.2	263	569	46.2	242	530	45.7	99	517	19.1

Note: N = Numerator; D= Denominator.

1. Heartland/Southwest, Midwest, and Pacific RCCs included in aggregate for Q2 2020 Measure 4 Immunization Status. The SE region does not collect immunizations (adult or pediatric). NE provides data on a 6-month schedule.

*Vaccine priority for this project

Aggregation Across Quarters: Q1 2019 – Q2 2020

The aggregated data more thoroughly represents a population level view of the data across the nation. In order to compare across RCCs, NE data for Q2 2019 (Combined Q1/Q2: January 1, 2019 – June 30, 2019), Q4 2019 (Combined Q3/Q4: July 1, 2019 – December 31, 2019), and Q2 2020 (Combined Q1/Q2: January 1, 2020 – June 30, 2020) are included for each aggregation.

On page 6, you will find the most recent (Q1/Q2 2020) data submission in cross quarter aggregation. For a visual representation of the aggregation across quarters, see graphs starting on page 8.

Aggregation Across Quarters: Q1 – Q2 2019 (January 1-June 30, 2019)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	5249	7526	69.7	586	3548	16.5	3348	5076	66.0	434	986	44.0	568
Adult	2500	4389	57.0	679	3463	19.6							

QI Measure 4 (Immunization Status)																					
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3082	3970	77.6	3111	3770	82.5	2406	3977	60.5	3125	3989	78.3	2202	3954	55.7	1819	3664	49.6	797	3693	21.6
Adult	1189	2066	57.6	1271	2129	59.7	1103	1952	56.5	1287	2304	55.9	742	1950	38.1	371	1918	19.3	419	1889	22.2

Aggregation Across Quarters: Q3 – Q4 2019 (July 1-December 31, 2019)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	7415	10436	71.1	989	7377	13.4	4139	6129	67.5	710	1860	38.2	366
Adult	3731	6142	60.7	1200	5267	22.8							

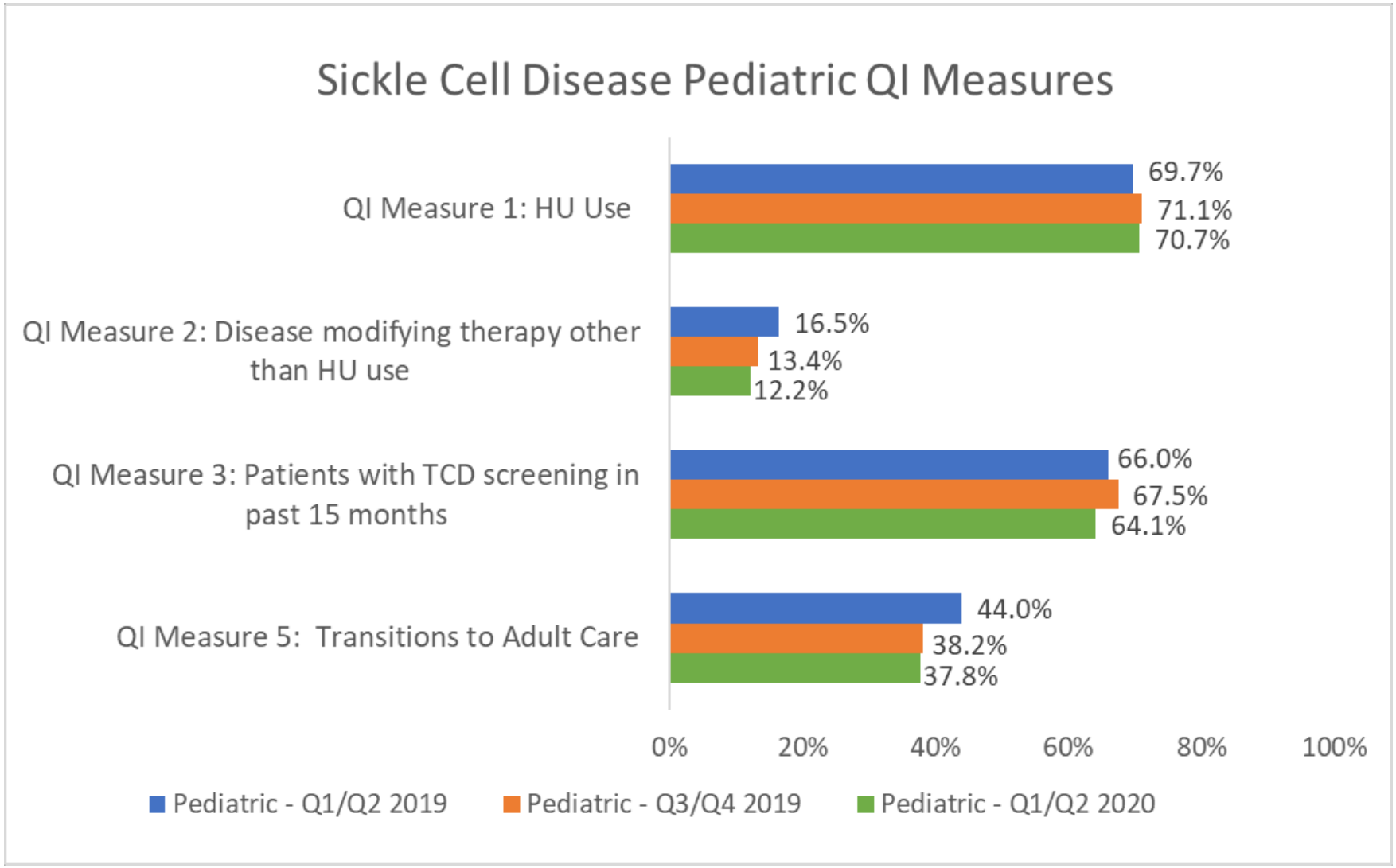
QI Measure 4 (Immunization Status)																				
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3450	4247	81.2	3507	4235	82.8	2770	4234	65.4	3213	4215	76.2	2017	4247	47.5	2394	3429	69.8	1068	3434	31.1
Adult	992	1653	60.0	1120	1711	65.5	769	1520	50.6	850	1470	57.8	662	1520	43.6	426	1100	38.7	182	1072	17.6

Aggregation Across Quarters: Q1 – Q2 2020 (January 1-June 30, 2020)

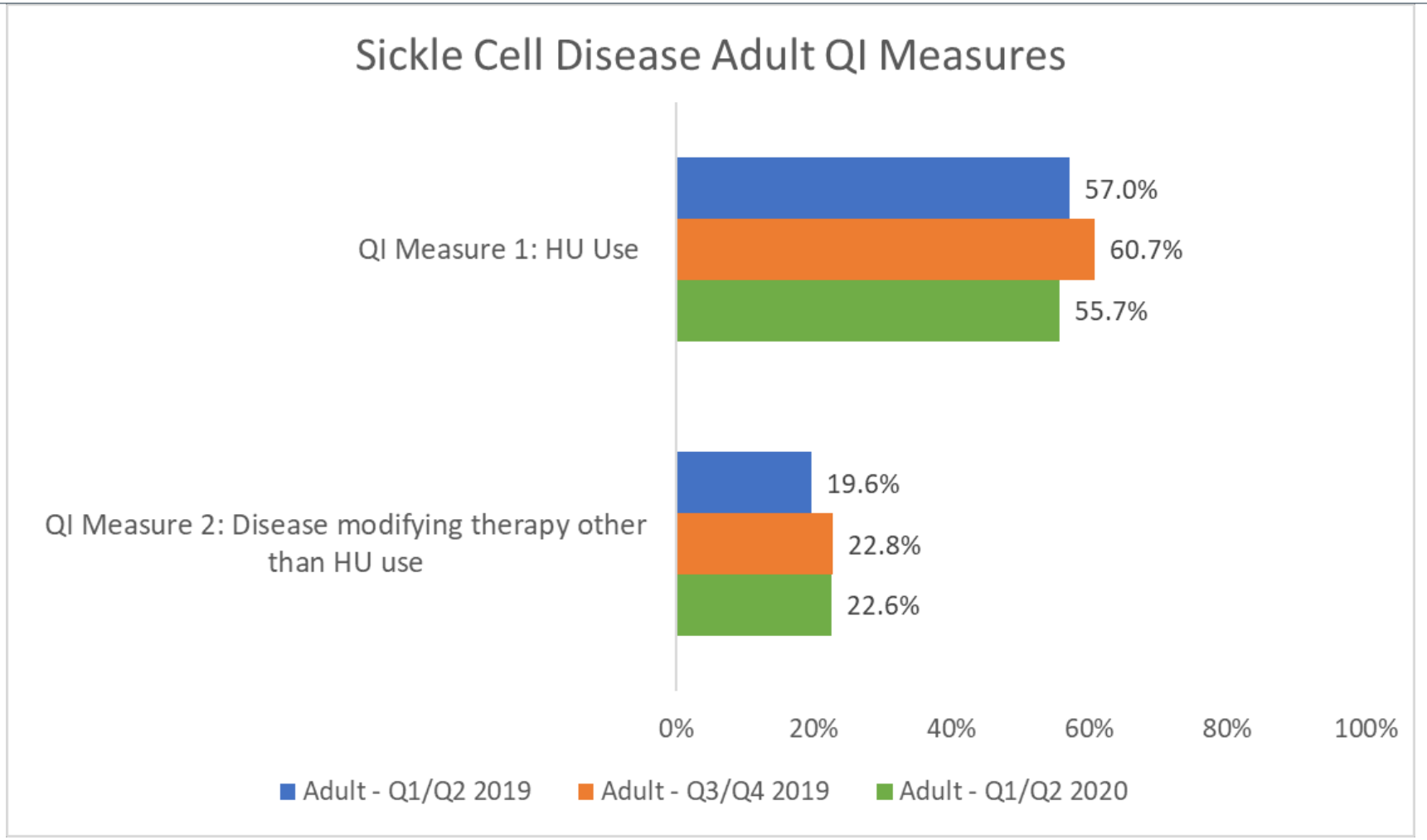
	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	7987	11298	70.7	930	7603	12.2	4654	7262	64.1	804	2129	37.8	1534
Adult	3560	6390	55.7	1269	5623	22.6							

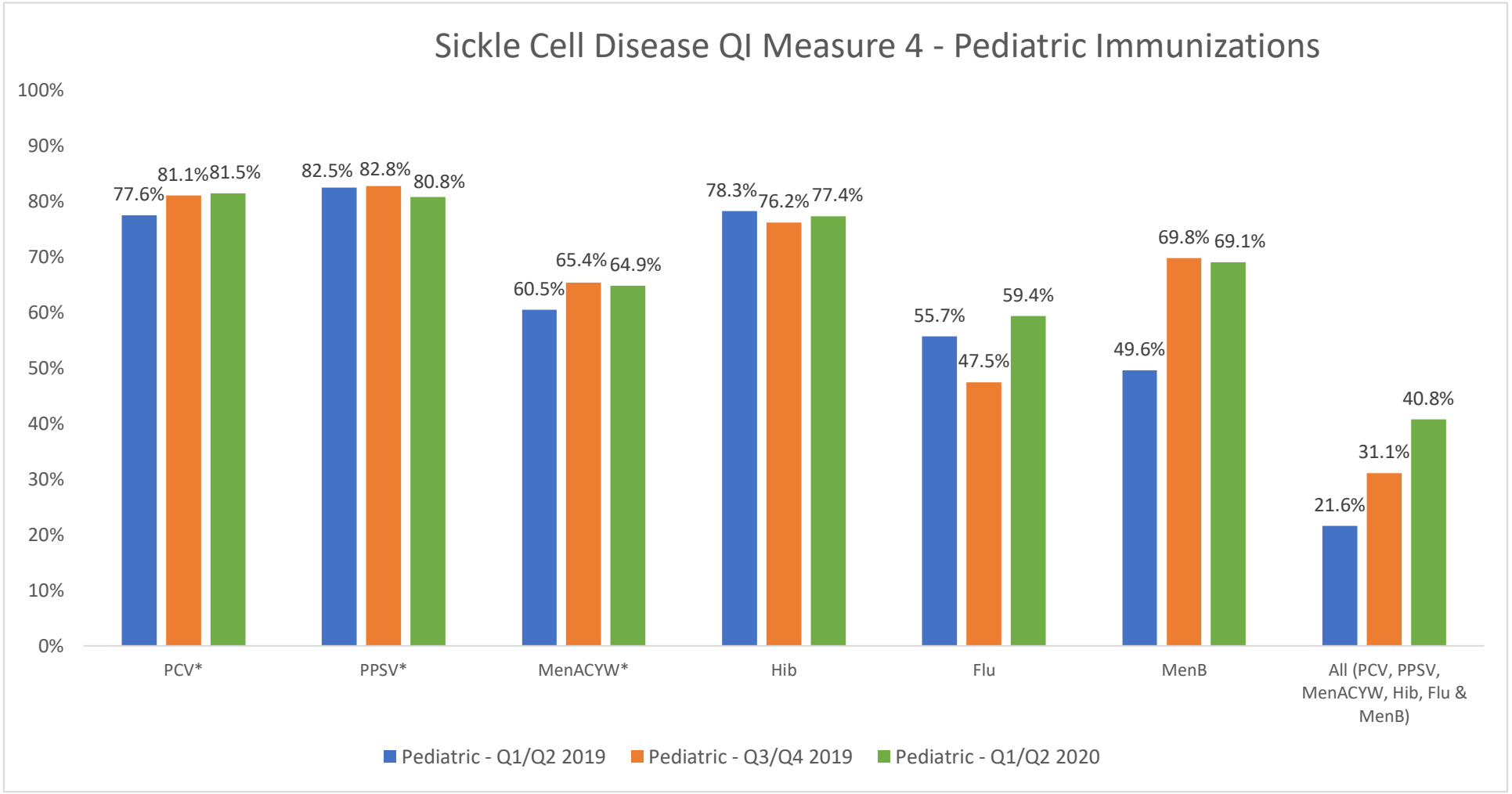
QI Measure 4 (Immunization Status)																				
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3645	4470	81.5	3367	4167	80.8	2711	4175	64.9	3323	4293	77.4	2639	4444	59.4	1798	2601	69.1	1342	3287	40.8
Adult	1422	2238	63.5	1501	2305	65.1	1263	2099	60.2	947	1570	60.3	841	1743	48.3	966	1647	58.7	429	1462	29.3

Graphs of Aggregation Across Quarters



8


Sickle Cell Disease Adult QI Measures
QI Measure 1: HU Use
57.0%
60.7%
55.7%
QI Measure 2: Disease modifying therapy other than HU use
19.6%
22.8%
22.6%
0%
20%
40%
60%
80%
100%
Adult - Q1/Q2 2019
Adult - Q3/Q4 2019
Adult - Q1/Q2 2020


Sickle Cell Disease QI Measure 4 - Pediatric Immunizations
100%
90%
80%
70%
60%
50%
40%
30%
20%
10%
0%
77.6%
81.1%
81.5%
82.5%
82.8%
80.8%
60.5%
65.4%
64.9%
78.3%
76.2%
77.4%
55.7%
47.5%
59.4%
49.6%
69.8%
69.1%
21.6%
31.1%
40.8%
PCV*
PPSV*
MenACYW*
Hib
Flu
MenB
All (PCV, PPSV, MenACYW, Hib, Flu & MenB)
Pediatric - Q1/Q2 2019
Pediatric - Q3/Q4 2019
Pediatric - Q1/Q2 2020

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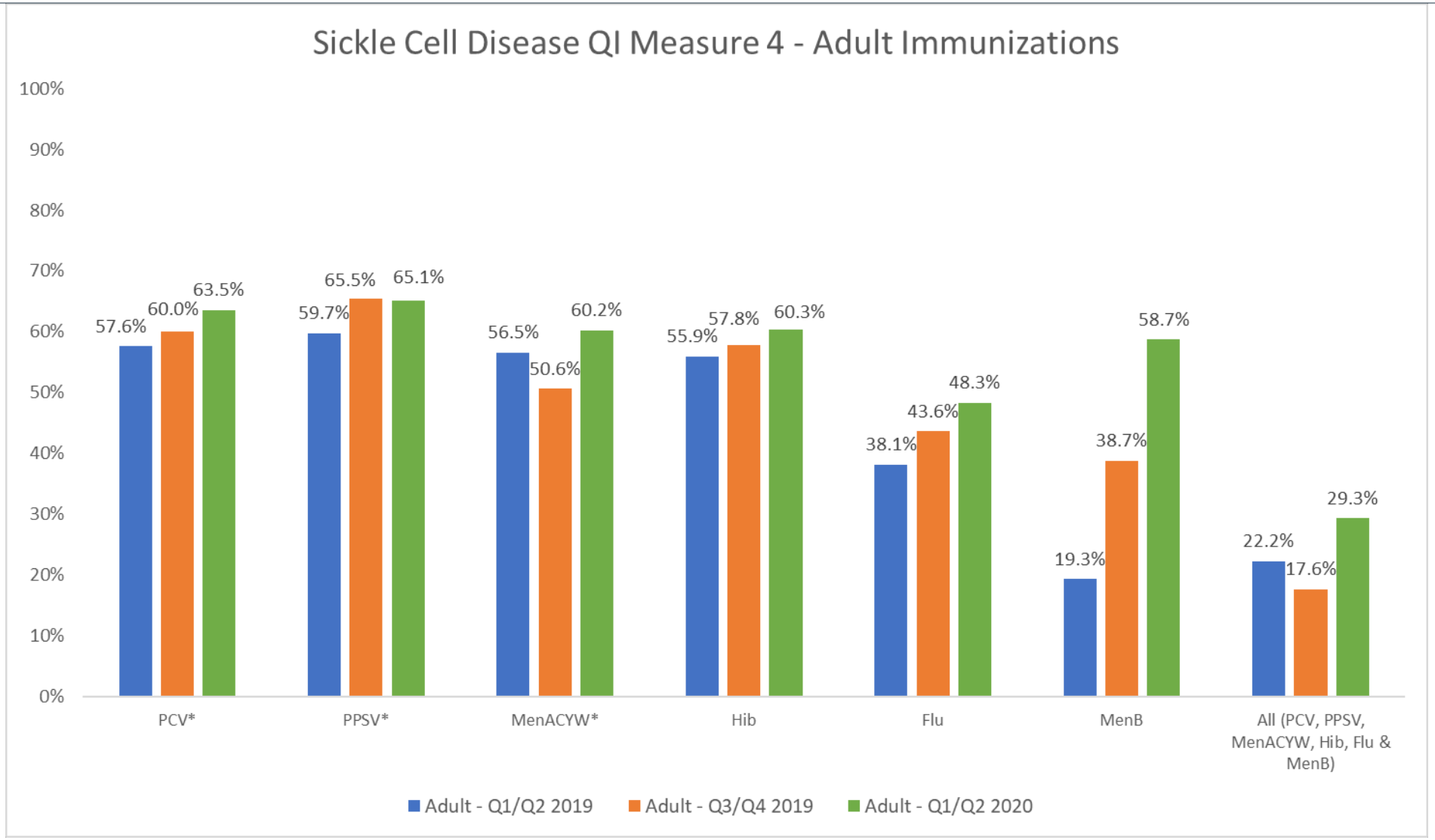
Sickle Cell Disease QI Measure 4 - Adult Immunizations
100%
90%
80%
70%
60%
50%
40%
30%
20%
10%
0%
57.6%
60.0%
63.5%
59.7%
65.5%
65.1%
56.5%
50.6%
60.2%
55.9%
57.8%
60.3%
38.1%
43.6%
48.3%
19.3%
38.7%
58.7%
22.2%
17.6%
29.3%
PCV*
PPSV*
MenACYW*
Hib
Flu
MenB
All (PCV, PPSV, MenACYW, Hib, Flu & MenB)
Adult - Q1/Q2 2019
Adult - Q3/Q4 2019
Adult - Q1/Q2 2020

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Region Specific Values for QI Measures

QI Measure 1a: Hydroxyurea use among Children/Adolescents (Required measure for all RCCs to collect)

Percent of patients >9-months and <18 years of age prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020		
	N	D	%	N	N	D	%	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	828	1144	72.4	849	1161	73.1	856	1153	74.2	904	1257	71.9	927	1258	73.7	957	1298	73.7
Midwest	345	437	78.9	338	393	86.0	418	537	77.8	361	439	82.2	413	508	81.3	526	645	81.6
Northeast[1]	643	883	72.8	775	1179	65.7				429	597	71.9				825	1159	71.2
Southeast				1552	2300	67.5	1910	2734	69.9	1958	2735	71.6	1972	2877	68.5	1898	2771	68.5
Pacific	277	453	61.2	285	459	62.1	288	468	61.5	291	516	56.4	206	369	55.8	263	413	63.7

1. Note, Northeast Q1 data included September 1, 2018 – February 28th, 2019. In order to compare across all RCCs, NE data for Q2 (Combined Q1/Q2: January 1, 2019 – June 30, 2019) and Q4 (Combined Q3/Q4: July 1, 2019 – December 31, 2019) were used as proxy for the data presented for across quarters. This footnote will not be repeated for each measure, however, is applicable across all.

QI Measure 1b: Hydroxyurea use among Adults (Required measure for all RCCs to collect)

Percent of patients 18 years and older prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	258	426	60.6	246	446	55.2	234	428	54.7	249	468	53.2	256	469	54.6	262	473	55.4
Midwest	75	108	69.4	90	119	75.6	96	119	80.7	93	119	78.2	84	111	75.7	88	109	80.7
Northeast	485	874	55.5	437	891	48.7				541	870	62.2				746	1257	59.4
Southeast				1157	1975	58.6	1151	1826	63.0	1156	1876	61.6	828	1605	51.6	1060	1874	56.6
Pacific	128	216	59.3	109	208	52.4	99	208	47.6	112	228	49.1	109	231	47.2	127	261	48.7

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents (Optional Measure)

Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	100	402	24.9	95	388	24.5	96	374	25.7	94	413	22.8	91	408	22.3	87	393	22.1
Midwest	54	413	13.1	58	540	10.7	58	482	12.0	86	539	16.0	23	198	11.6	59	571	10.3
Northeast	62	275	22.5	51	273	18.7				26	197	13.2				39	678	5.8
Southeast				204	1402	14.6	255	2359	10.8	350	2864	12.2	303	2742	11.1	328	2613	12.6
Pacific	12	65	18.5	12	65	18.5	12	67	17.9	12	82	14.6						

QI Measure 2b: Disease modifying therapy use other than HU among Adults (Optional Measure)

Percent of patients 18 years and older prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020		
	N	D	%	N	D	%	%	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	73	261	28.0	92	286	32.2	104	292	35.6	112	322	34.8	123	342	36.0	128	332	38.6
Midwest	21	104	20.2	12	105	11.4	13	105	12.4	20	103	19.4	18	83	21.7	12	92	13.0
Northeast	161	406	39.7	154	576	26.7				165	497	33.2				290	923	31.4
Southeast				318	2106	15.1	356	1823	19.5	424	2099	20.2	325	1785	18.2	373	2066	18.1
Pacific	4	12	33.3	5	13	38.5	4	16	25.0	2	10	20.0						

QI Measure 3: Transcranial Doppler (Optional Measure)

Eligible SCD patients between ages of 2-16, that had a Transcranial Doppler (TCD) screening within the 15 months prior to the end of the quarter

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	695	1041	66.8	722	1042	69.3	728	1031	70.6	777	1137	68.3	773	1144	67.6	769	1123	68.5
Midwest	299	371	80.6	389	477	81.6	393	472	83.3	415	523	79.3	358	442	81.0	451	578	78.0
Northeast	393	881	44.6	295	584	50.5				300	541	55.5				690	1463	47.2
Southeast				519	972	53.4	540	920	58.7	540	861	62.7	561	832	67.4	609	991	61.5
Pacific	214	283	75.6	215	306	70.3	222	307	72.3	224	337	66.5	207	325	63.7	236	364	64.8

QI Measure 4a: Immunization - Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[3]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8
Northeast	598	1493	40.1	754	1493	50.5	448	1493	30.0	525	1493	35.2	316	1493	21.2	38	1493	2.5	119	1493	8.0
Southeast																					
Pacific	18	29	62.1	17	29	58.6	20	29	69.0	6	29	20.7	16	29	55.2	13	29	44.8			

1. UTD= Up to date. This footnote will not be repeated in future tables.
2. Detailed information to match flu season with quarter were provided during Q2 2019. This footnote will not be repeated in future tables.
3. Region where estimates may not be accurate due to need to clarify how to report data if not all sites reported immunization data. Issue has since been resolved.

Quarter 2 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9
Northeast	815	1024	79.6	854	1024	83.4	620	1024	60.5	818	1024	79.9	344	1024	33.6	197	1024	19.2	182	1024	17.8
Southeast																					
Pacific	14	21	66.7	11	21	52.4	15	21	71.4	3	21	14.3	17	21	81.0	10	21	47.6			

Quarter 3 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	812	997	81.4	843	997	84.6	596	997	59.8	858	997	86.1	347	997	34.8	531	997	53.3	121	997	12.1
Midwest	534	630	84.8	494	630	78.4	501	630	79.5	428	630	67.9	361	630	57.3	487	630	77.3	216	630	34.3
Northeast																					
Southeast																					
Pacific	12	21	57.1	12	21	57.1	14	21	66.7	5	21	23.8	13	21	61.9	10	12	83.3			

Quarter 4 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	919	1080	85.1	942	1080	87.2	720	1080	66.7	949	1080	87.9	518	1080	48.0	598	855	69.9	227	855	26.6
Midwest	600	661	90.8	528	656	80.5	523	652	80.2	496	661	75.0	456	661	69.0	580	648	89.5	297	661	44.9
Northeast	554	834	66.4	617	827	81.1	399	830	48.1	463	802	57.7	307	834	36.8	171	263	65.0	207	291	71.1
Southeast																					
Pacific	19	24	79.2	17	24	70.8	17	24	70.8	14	24	58.3	15	24	62.5	17	24	70.8			

Quarter 1 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	934	1102	84.8	944	1102	85.7	722	1102	65.5	970	1102	88.0	646	1102	58.6	618	876	70.5	341	876	38.9
Midwest	594	650	91.4	397	503	78.9	396	526	75.3	430	594	72.4	461	657	70.2	147	217	67.7	300	672	44.6
Northeast																					
Southeast																					
Pacific	19	21	90.5	17	21	81.0	18	21	85.7	15	21	71.4	16	21	76.2	18	21	85.7			

Quarter 2 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	926	1081	85.7	949	1081	87.8	786	1081	72.7	955	1081	88.3	646	1081	59.8	695	857	81.1	336	857	39.2
Midwest	572	617	92.7	367	461	79.6	388	510	76.1	409	560	73.0	410	603	68.0	135	200	67.5	279	630	44.3
Northeast	585	982	59.6	679	982	69.1	387	918	42.2	533	918	58.1	446	963	46.3	171	413	41.4	86	252	34.1
Southeast																					
Pacific	15	17	88.2	14	17	82.4	14	17	82.4	11	17	64.7	14	17	82.4	14	17	82.4			

QI Measure 4b: Immunization - Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[1]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	151	295	51.2	20	263	7.60	13	263	4.94
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.2
Southeast																					
Pacific	11	94	11.7	35	111	31.5	10	17	58.8	142	191	74.3	10	17	58.8	7	17	41.2			

1. Region where estimates may not be accurate due to need to clarify how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5
Northeast	625	933	67.0	525	933	56.3	511	933	54.8	485	933	52.0	186	933	19.9	36	933	3.9	270	933	28.9
Southeast																					
Pacific	21	83	25.3	38	95	40.0	5	12	41.7	141	190	74.2	6	12	50.0	4	12	33.3			

Quarter 3 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	184	264	69.7	200	264	75.8	153	264	58.0	159	264	60.2	122	264	46.2	59	264	22.3	31	264	11.7
Midwest	170	212	80.2	140	212	66.0	148	212	69.8	100	212	47.2	131	212	61.8	125	212	59.0	58	212	27.4
Northeast																					
Southeast																					
Pacific	23	79	29.1	39	91	42.9	4	12	33.3	125	182	68.7	6	12	50.0	3	12	25.0			

Quarter 4 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	172	295	58.3	225	295	76.3	162	295	54.9	157	295	53.2	116	295	39.3	55	263	20.9	28	263	10.6
Midwest	179	213	84.0	151	213	70.9	157	213	73.7	110	213	51.6	91	213	42.7	137	213	64.3	45	213	21.1
Northeast	238	508	46.9	320	538	59.5	142	508	28.0	55	120	45.8	189	508	37.2	41	120	34.2	27	120	22.5
Southeast																					
Pacific	26	82	31.7	45	98	45.9	3	16	18.8	144	184	78.3	7	16	43.8	6	16	37.5			

Quarter 1 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	201	310	64.8	219	310	70.6	156	310	50.3	176	310	56.8	151	310	48.7	68	271	25.1	44	271	16.2
Midwest	176	214	82.2	150	214	70.1	156	214	72.9	110	214	51.4	93	214	43.5	138	214	64.5	44	214	20.6
Northeast																					
Southeast																					
Pacific	27	82	32.9	50	99	50.5	6	17	35.3	149	188	79.3	9	17	52.9	5	17	29.4			

Quarter 2 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	221	323	68.4	238	323	73.7	177	323	54.8	185	323	57.3	164	323	50.8	87	284	30.6	57	284	20.1
Midwest	194	233	83.3	154	233	66.1	155	233	66.5	107	233	49.9	92	233	39.5	152	233	65.2	42	233	18.0
Northeast	576	989	58.2	645	1026	62.9	607	989	61.4	56	125	44.8	325	633	51.3	513	615	83.4	242	460	52.6
Southeast																					
Pacific	27	87	31.0	45	100	45.0	6	13	46.2	164	177	92.7	7	13	53.8	3	13	23.1			

QI Measure 5: Transitions to Adult Care - Number of patients that have a documented transition education discussion

	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020		
Region	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Heartland/Southwest	81	165	49.1	93	161	57.8	117	184	63.6	118	181	65.2	108	173	62.4	109	183	59.6
Midwest	9	48	18.8	21	63	33.3	16	63	25.4	18	59	30.5	20	70	28.8	15	63	23.8
Northeast	21	110	19.1	68	175	38.9				6	91	6.6				66	275	24.0
Pacific	16	23	69.6	14	22	63.6	13	19	68.4	11	18	61.1	12	17	70.6	8	13	61.5
Southeast				132	329	40.1	186	562	33.1	225	683	32.9	226	686	32.9	240	649	37.0

QI Measure 6: ECHO - Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Q1 2019	Q2 2019	Q3 2019	Q4 2019	Q1 2020	Q2 2020
Heartland/Southwest	61	43	58	54	35	87
Midwest	48	49	35	46	120	451
Northeast	170	242		146		580
Pacific	59		52	65	88	68
Southeast		66	56	55	72	33

Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP)

Quality Improvement Data Summary

Quarter 3 2020

January 8, 2021

Prepared by:

NICHQ's Department of Applied Research and Evaluation (DARE)

National Coordinating Center for the SCDTDRCP

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Executive Summary

To address the ongoing challenges of serving children and adults with sickle cell disease (SCD), and to improve care and outcomes, Congress created legislation to fund the SCDTDRCP. The goals of the SCDTDRCP are to 1) improve coordination and service delivery for individuals living with SCD; 2) improve access to services; and 3) improve and expand on provider knowledge of SCD treatment and care. The SCDTDRCP funds five Regional Coordinating Centers (RCCs) and one National Coordinating Center (NCC).

RCCs are responsible for quarterly reporting on at least two Quality Improvement (QI) Measures. All regions must assess Hydroxyurea (HU) use; the regions can choose which other QI measure(s) to collect. The six QI measures include: 1) Hydroxyurea use; 2) Other disease modifying therapy use; 3) Immunization status; 4) Transcranial Doppler Ultrasound screening; 5) Transition to adult care; and 6) Providers' participation in Extension of Community Healthcare Outcomes (ECHO).

Data in this report includes: 1) aggregate data from all regions reporting for the quarter and 2) regional data by quarter.

As of Quarter 3 2020, the RCCs have been collecting data for seven quarters. Below are observations from the data collection to date:

- RCCs, which are centers of excellence, and the local sites within the regions, are making strides towards consistent in implementation of guideline-based care for SCD.
- RCCs consistently capture: Prescribing of HU and TCD screening, which indicates necessary infrastructure at local site and RCC regarding the collect these data.
- ECHO has become a standard practice for all regions, and attendance for these sessions have been consistently tracked.
- RCCs report that capturing complete immunization data has been challenging. Consistency and volume of submitted data is variable.
- RCCs confirm that transitional care is important and sites that have selected this measure have had success in collecting required data. However, comparison between sites is limited since the definition of transition care is inconsistent between sites.

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Overview: This report reflects data submitted for Quarter 3 2020 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP) which spanned July 1 to September 30, 2020. Regional Coordinating Centers (RCCs) aggregated and submitted data to CoLab between October 1 and November 15, 2020.

Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers' participation in ECHO

Regions collected data for this QI report via reporting from Electronic Health Records (EHR) and/or by manual chart review at the site level. For Quarter 3 2020, four of the regions submitted quarterly data (Heartland/Southwest, Midwest, Pacific and Southeast). The Northeast RCC provides data on a 6-month schedule (reporting for Q2, Q4). All data is re-run on a 6-month basis (at Q2 and Q4) and any updates made to previous quarterly data are reflected at these intervals. The last re-run of all data occurred on **8/19/20**.

The first page of this report displays the aggregated values for the QI measures across regions for Q3 2020. With the remainder of the report consisting of QI measures by region.

The sample for each region reflects the total number of sickle-cell patients seen from participating sites of a region within the specific quarter. Greyed out boxes indicate that an RCC did not provide data either because it was not applicable or not collected. Notes about annotations are provided where relevant. Headings for the current Q3 2020 data are highlighted in green to assist in readability across quarters. Guidance to RCCs about inclusion for numerators and denominators is detailed in the Manual of Operating Procedures (MOP). Linked here is the most up-to-date MOP.

Acknowledgement of the potential impact of COVID-19: Starting in March 2020, the COVID-19 Pandemic was present in the United States, enough to disrupt the usual cadence of life. Many of the SCDTDRCP providers at the local and regional levels were engaged in either front-line care or planning and developing new procedures and processes to respond to emerging needs. All SCDTDRCP regions reported upheaval in their clinical systems and concern that appointments, both elective and/or essential, were not occurring for SCD patients as usual or as recommended. Therefore, there is some known (i.e., certain sites have been unable to report data due to staff reductions) and potentially unknown (i.e., reduced or eliminated visits, decisions not to change medication during this time, clinical priority shifts) variation in 2020 data. As numbers are reported in 2020 and beyond, it will be important to read them in the context of their collection occurring during the COVID-19 pandemic.

Aggregated Values for Q3 2020 QI Measures (excluding QI Measure 4, immunizations- see below)[1]

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[2]			QI Measure 5: Transitions to Adult Care[3]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	3508	4909	71.5	461	3191	14.5	2028	3100	65.4	368	842	43.7	424
Adult	1739	2652	65.6	929	2202	42.2							

Note: N = Numerator; D= Denominator

1. Data included in aggregate values for Q3 2020 includes four regions (Heartland/Southwest, Midwest, Pacific and Southeast).
2. Only for eligible patients between 2 and 16 years of age
3. Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q3 2020 QI Measure 4 (Immunization Status)[1]

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	1535	1755	87.5	1383	1611	85.9	1226	1629	75.3	1561	1867	83.6	676	1758	38.5	850	1072	79.3	420	1538	27.3
Adult	450	639	70.4	444	639	69.5	345	557	61.9	458	736	62.2	172	557	30.9	249	513	48.5	74	513	14.4

Note: N = Numerator; D= Denominator.

1. Heartland/Southwest, Midwest, and Pacific RCCs included in aggregate for Q3 2020 Measure 4 Immunization Status. The SE region does not collect immunizations (adult or pediatric). NE provides data on a 6-month schedule.

*Vaccine priority for this project

QI Measure 1a: Hydroxyurea use among Children/Adolescents (Required measure for all RCCs to collect)

Percent of patients >9-months and <18 years of age prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020		
	N	D	%	N	N	D	%	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	828	1144	72.4	849	1161	73.1	856	1153	74.2	904	1257	71.9	927	1258	73.7	957	1298	73.7	955	1298	73.6
Midwest	345	437	78.9	338	393	86.0	418	537	77.8	361	439	82.2	413	508	81.3	526	645	81.6	423	518	81.7
Northeast[1]	643	883	72.8	775	1179	65.7				429	597	71.9				825	1159	71.2			
Southeast				1552	2300	67.5	1910	2734	69.9	1958	2735	71.6	1972	2877	68.5	1898	2771	68.5	1878	2669	70.4
Pacific	277	453	61.2	285	459	62.1	288	468	61.5	291	516	56.4	206	369	55.8	263	413	63.7	252	424	59.4

1. Note, Northeast Q1 data included September 1, 2018 – February 28th, 2019. In order to compare across all RCCs, NE data for Q2 (Combined Q1/Q2: January 1, 2019 – June 30, 2019) and Q4 (Combined Q3/Q4: July 1, 2019 – December 31, 2019) were used as proxy for the data presented for across quarters. This footnote will not be repeated for each measure, however, is applicable across all.

QI Measure 1b: Hydroxyurea use among Adults (Required measure for all RCCs to collect)

Percent of patients 18 years and older prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	258	426	60.6	246	446	55.2	234	428	54.7	249	468	53.2	256	469	54.6	262	473	55.4	246	438	56.2
Midwest	75	108	69.4	90	119	75.6	96	119	80.7	93	119	78.2	84	111	75.7	88	109	80.7	90	116	77.6
Northeast	485	874	55.5	437	891	48.7				541	870	62.2				746	1257	59.4			
Southeast				1157	1975	58.6	1151	1826	63.0	1156	1876	61.6	828	1605	51.6	1060	1874	56.6	1293	1843	70.2
Pacific	128	216	59.3	109	208	52.4	99	208	47.6	112	228	49.1	109	231	47.2	127	261	48.7	110	255	43.1

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents (Optional Measure)

Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	100	402	24.9	95	388	24.5	96	374	25.7	94	413	22.8	91	408	22.3	87	393	22.1	100	401	24.9
Midwest	54	413	13.1	58	540	10.7	58	482	12.0	86	539	16.0	23	198	11.6	59	571	10.3	51	498	10.2
Northeast	62	275	22.5	51	273	18.7				26	197	13.2				39	678	5.8			
Southeast				204	1402	14.6	255	2359	10.8	350	2864	12.2	303	2742	11.1	328	2613	12.6	310	2292	13.5
Pacific	12	65	18.5	12	65	18.5	12	67	17.9	12	82	14.6									

QI Measure 2b: Disease modifying therapy use other than HU among Adults (Optional Measure)

Percent of patients 18 years and older prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020		
	N	D	%	N	D	%	%	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	73	261	28.0	92	286	32.2	104	292	35.6	112	322	34.8	123	342	36.0	128	332	38.6	131	314	41.7
Midwest	21	104	20.2	12	105	11.4	13	105	12.4	20	103	19.4	18	83	21.7	12	92	13.0	12	99	12.1
Northeast	161	406	39.7	154	576	26.7				165	497	33.2				290	923	31.4			
Southeast				318	2106	15.1	356	1823	19.5	424	2099	20.2	325	1785	18.2	373	2066	18.1	786	1789	43.9
Pacific	4	12	33.3	5	13	38.5	4	16	25.0	2	10	20.0									

QI Measure 3: Transcranial Doppler (Optional Measure)

Eligible SCD patients between ages of 2-16, that had a Transcranial Doppler (TCD) screening within the 15 months prior to the end of the quarter

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020		
	N	D	%	N	D	%	%	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	695	1041	66.8	722	1042	69.3	728	1031	70.6	777	1137	68.3	773	1144	67.6	769	1123	68.5	753	1155	65.2
Midwest	299	371	80.6	389	477	81.6	393	472	83.3	415	523	79.3	358	442	81.0	451	578	78.0	445	570	78.1
Northeast	393	881	44.6	295	584	50.5				300	541	55.5				690	1463	47.2			
Southeast				519	972	53.4	540	920	58.7	540	861	62.7	561	832	67.4	609	991	61.5	566	980	57.8
Pacific	214	283	75.6	215	306	70.3	222	307	72.3	224	337	66.5	207	325	63.7	236	364	64.8	264	395	66.8

QI Measure 4a: Immunization - Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[3]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8
Northeast	598	1493	40.1	754	1493	50.5	448	1493	30.0	525	1493	35.2	316	1493	21.2	38	1493	2.5	119	1493	8.0
Southeast																					
Pacific	18	29	62.1	17	29	58.6	20	29	69.0	6	29	20.7	16	29	55.2	13	29	44.8			

1. UTD= Up to date. This footnote will not be repeated in future tables.
2. Detailed information to match flu season with quarter were provided during Q2 2019. This footnote will not be repeated in future tables.
3. Region where estimates may not be accurate due to need to clarify how to report data if not all sites reported immunization data. Issue has since been resolved.

Quarter 2 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9
Northeast	815	1024	79.6	854	1024	83.4	620	1024	60.5	818	1024	79.9	344	1024	33.6	197	1024	19.2	182	1024	17.8
Southeast																					
Pacific	14	21	66.7	11	21	52.4	15	21	71.4	3	21	14.3	17	21	81.0	10	21	47.6			

Quarter 3 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	812	997	81.4	843	997	84.6	596	997	59.8	858	997	86.1	347	997	34.8	531	997	53.3	121	997	12.1
Midwest	534	630	84.8	494	630	78.4	501	630	79.5	428	630	67.9	361	630	57.3	487	630	77.3	216	630	34.3
Northeast																					
Southeast																					
Pacific	12	21	57.1	12	21	57.1	14	21	66.7	5	21	23.8	13	21	61.9	10	12	83.3			

Quarter 4 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	919	1080	85.1	942	1080	87.2	720	1080	66.7	949	1080	87.9	518	1080	48.0	598	855	69.9	227	855	26.6
Midwest	600	661	90.8	528	656	80.5	523	652	80.2	496	661	75.0	456	661	69.0	580	648	89.5	297	661	44.9
Northeast	554	834	66.4	617	827	81.1	399	830	48.1	463	802	57.7	307	834	36.8	171	263	65.0	207	291	71.1
Southeast																					
Pacific	19	24	79.2	17	24	70.8	17	24	70.8	14	24	58.3	15	24	62.5	17	24	70.8			

Quarter 1 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	934	1102	84.8	944	1102	85.7	722	1102	65.5	970	1102	88.0	646	1102	58.6	618	876	70.5	341	876	38.9
Midwest	594	650	91.4	397	503	78.9	396	526	75.3	430	594	72.4	461	657	70.2	147	217	67.7	300	672	44.6
Northeast																					
Southeast																					
Pacific	19	21	90.5	17	21	81.0	18	21	85.7	15	21	71.4	16	21	76.2	18	21	85.7			

Quarter 2 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	926	1081	85.7	949	1081	87.8	786	1081	72.7	955	1081	88.3	646	1081	59.8	695	857	81.1	336	857	39.2
Midwest	572	617	92.7	367	461	79.6	388	510	76.1	409	560	73.0	410	603	68.0	135	200	67.5	279	630	44.3
Northeast	585	982	59.6	679	982	69.1	387	918	42.2	533	918	58.1	446	963	46.3	171	413	41.4	86	252	34.1
Southeast																					
Pacific	15	17	88.2	14	17	82.4	14	17	82.4	11	17	64.7	14	17	82.4	14	17	82.4			

Quarter 3 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	945	1091	86.6	971	1091	89.0	824	1091	75.5	970	1091	88.9	268	1091	24.6	710	861	82.5	145	861	16.8
Midwest	590	664	88.9	412	520	79.2	402	538	74.7	446	608	73.4	408	667	61.2	140	211	66.4	275	677	40.6
Northeast																					
Southeast																					
Pacific[1]										145	168	86.3									

1. Additional data forthcoming

QI Measure 4b: Immunization - Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[1]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	151	295	51.2	20	263	7.60	13	263	4.94
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.64	184	2009	9.2
Southeast																					
Pacific	11	94	11.7	35	111	31.5	10	17	58.8	142	191	74.3	10	17	58.8	7	17	41.2			

1. Region where estimates may not be accurate due to need to clarify how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5
Northeast	625	933	67.0	525	933	56.3	511	933	54.8	485	933	52.0	186	933	19.9	36	933	3.9	270	933	28.9
Southeast																					
Pacific	21	83	25.3	38	95	40.0	5	12	41.7	141	190	74.2	6	12	50.0	4	12	33.3			

Quarter 3 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	184	264	69.7	200	264	75.8	153	264	58.0	159	264	60.2	122	264	46.2	59	264	22.3	31	264	11.7
Midwest	170	212	80.2	140	212	66.0	148	212	69.8	100	212	47.2	131	212	61.8	125	212	59.0	58	212	27.4
Northeast																					
Southeast																					
Pacific	23	79	29.1	39	91	42.9	4	12	33.3	125	182	68.7	6	12	50.0	3	12	25.0			

Quarter 4 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	172	295	58.3	225	295	76.3	162	295	54.9	157	295	53.2	116	295	39.3	55	263	20.9	28	263	10.6
Midwest	179	213	84.0	151	213	70.9	157	213	73.7	110	213	51.6	91	213	42.7	137	213	64.3	45	213	21.1
Northeast	238	508	46.9	320	538	59.5	142	508	28.0	55	120	45.8	189	508	37.2	41	120	34.2	27	120	22.5
Southeast																					
Pacific	26	82	31.7	45	98	45.9	3	16	18.8	144	184	78.3	7	16	43.8	6	16	37.5			

Quarter 1 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	201	310	64.8	219	310	70.6	156	310	50.3	176	310	56.8	151	310	48.7	68	271	25.1	44	271	16.2
Midwest	176	214	82.2	150	214	70.1	156	214	72.9	110	214	51.4	93	214	43.5	138	214	64.5	44	214	20.6
Northeast																					
Southeast																					
Pacific	27	82	32.9	50	99	50.5	6	17	35.3	149	188	79.3	9	17	52.9	5	17	29.4			

Quarter 2 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	221	323	68.4	238	323	73.7	177	323	54.8	185	323	57.3	164	323	50.8	87	284	30.6	57	284	20.1
Midwest	194	233	83.3	154	233	66.1	155	233	66.5	107	233	49.9	92	233	39.5	152	233	65.2	42	233	18.0
Northeast	576	989	58.2	645	1026	62.9	607	989	61.4	56	125	44.8	325	633	51.3	513	615	83.4	242	460	52.6
Southeast																					
Pacific	27	87	31.0	45	100	45.0	6	13	46.2	164	177	92.7	7	13	53.8	3	13	23.1			

Quarter 3 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	239	337	70.9	254	337	75.4	197	337	58.5	202	337	59.9	71	337	21.1	102	293	34.8	35	293	12.0
Midwest	183	220	83.2	152	220	69.1	148	220	67.3	111	220	50.5	101	220	45.9	147	220	66.8	39	220	17.7
Northeast																					
Southeast																					
Pacific	28	82	34.1	38	82	46.3				145	179	81.0									

QI Measure 5: Transitions to Adult Care - Number of patients that have a documented transition education discussion

	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020		
Region	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Heartland/Southwest	81	165	49.1	93	161	57.8	117	184	63.6	118	181	65.2	108	173	62.4	109	183	59.6	114	175	65.1
Midwest	9	48	18.8	21	63	33.3	16	63	25.4	18	59	30.5	20	70	28.8	15	63	23.8	19	71	26.8
Northeast	21	110	19.1	68	175	38.9				6	91	6.6				66	275	24.0			
Pacific	16	23	69.6	14	22	63.6	13	19	68.4	11	18	61.1	12	17	70.6	8	13	61.5	5	9	55.6
Southeast				132	329	40.1	186	562	33.1	225	683	32.9	226	686	32.9	240	649	37.0	230	587	39.2

QI Measure 6: ECHO - Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Q1 2019	Q2 2019	Q3 2019	Q4 2019	Q1 2020	Q2 2020	Q3 2020
Heartland/Southwest	61	43	58	54	35	87	64
Midwest	48	49	35	46	120	451	235
Northeast	170	242		146		580	
Pacific	59		52	65	88	68	52
Southeast		66	56	55	72	33	73

Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP)

Quality Improvement Data Summary

Quarter 4 2020

March 18, 2021

Prepared by:

NICHQ's Department of Applied Research and Evaluation (DARE)

National Coordinating Center for the SCDTDRCP

Executive Summary

To address the ongoing challenges of serving children and adults with sickle cell disease (SCD), and to improve care and outcomes, Congress created legislation to fund the SCDTDRCP. The goals of the SCDTDRCP are to 1) improve coordination and service delivery for individuals living with SCD; 2) improve access to services; and 3) improve and expand on provider knowledge of SCD treatment and care. The SCDTDRCP funds five Regional Coordinating Centers (RCCs) and one National Coordinating Center (NCC).

RCCs are responsible for quarterly reporting on at least two Quality Improvement (QI) Measures. All regions must assess Hydroxyurea (HU) use; the regions can choose which other QI measure(s) to collect. The six QI measures include: 1) Hydroxyurea use; 2) Other disease modifying therapy use; 3) Immunization status; 4) Transcranial Doppler Ultrasound screening; 5) Transition to adult care; and 6) Providers' participation in Extension of Community Healthcare Outcomes (ECHO).

Data in this report includes aggregate data from all regions reporting for the quarter, aggregate data across multiple quarters to provide population level view, and regional data by quarter.

As of Quarter 4 2020, the RCCs have been collecting data for seven quarters. Below are observations from the data collection to date:

- RCCs, which are centers of excellence, and the local sites within the regions, are making strides towards consistent implementation of guideline-based care for SCD.
- RCCs consistently capture: Prescribing of HU and TCD screening, which indicates necessary infrastructure at local sites and RCCs regarding the collection of these data.
- ECHO has become a standard practice for all regions, and attendance for these sessions have been consistently tracked.
- RCCs report that capturing complete immunization data has been challenging. Consistency and volume of submitted data is variable.
- RCCs confirm that transitional care is important and sites that have selected this measure have had success in collecting required data. However, comparison between sites is limited since the definition of transition care is inconsistent between sites.

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Overview: This report reflects data submitted for Quarter 4 2020 QI data collection for the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP) which spanned October 1 to December 31, 2020. Regional Coordinating Centers (RCCs) aggregated and submitted data to CoLab between January 1 and February 15, 2021.

Data were intended to be a population-level view of key Quality Improvement (QI) metrics for all regions participating in SCDTDRCP:

1) Hydroxyurea use
2) Other disease modifying therapy use
3) Immunization status
4) Transcranial Doppler Ultrasound screening
5) Transitions to adult care
6) Providers' participation in ECHO

Regions collected data for this QI report via reporting from Electronic Health Records (EHR) and/or by manual chart review at the site level. For Quarter 4 2020, four of the regions submitted quarterly data (Heartland/Southwest, Midwest, Pacific and Southeast). The Northeast RCC provides data on a 6-month schedule (reporting for Q2, Q4). All data is re-run on a 6-month basis (at Q2 and Q4) and any updates made to previous quarterly data are reflected at these intervals. The last re-run of all data occurred on **2/17/21**.

Page five of this report displays the aggregated values for the QI measures across regions for Q4 2020 and then presents biannual aggregate values (i.e., combining Quarters 1 & 2 and Quarters 3 & 4) based on how frequently regional data was submitted. Starting on page six, these biannual aggregate values across quarters are included to provide a population-level perspective of QI measures. These are also displayed in graphs. Pages 15 through 23 provide QI measures by region.

The sample for each region reflects the total number of sickle-cell patients seen from participating sites of a region within the specific quarter. Greyed out boxes indicate that an RCC did not provide data either because it was not applicable or not collected. Notes about annotations are provided where relevant. Headings for the current Q4 2020 data are highlighted in green to assist in readability across quarters. Guidance to RCCs about inclusion for numerators and denominators is detailed in the Manual of Operating Procedures (MOP). Linked here is the most up-to-date MOP.

Acknowledgement of the potential impact of COVID-19: Starting in March 2020, the COVID-19 Pandemic was present in the United States, enough to disrupt the usual cadence of life. Many of the SCDTDRCP providers at the local and regional levels were engaged in either front-line care or planning and developing new procedures and processes to respond to emerging needs. All SCDTDRCP regions reported upheaval in their clinical systems and concern that appointments, both elective and/or essential, were not occurring for SCD patients as usual or as recommended. Therefore, there is some known (i.e., certain sites have been unable to report data due to staff reductions) and potentially unknown (i.e., reduced or eliminated visits, decisions not to change medication during this time, clinical priority shifts) variation in 2020 data. As numbers are reported in 2020 and beyond, it will be important to read them in the context of their collection occurring during the COVID-19 pandemic.

Aggregated Values for Q4 2020 QI Measures (excluding QI Measure 4, immunizations- see below)[1]

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months[2]			QI Measure 5: Transitions to Adult Care[3]			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	3522	4847	72.7	455	3192	14.3	2111	3122	67.6	345	827	41.7	212
Adult	1676	2936	57.1	661	2537	26.1							

Note: N = Numerator; D= Denominator

1. Data included in aggregate values for Q4 2020 includes four regions (Heartland/Southwest, Midwest, Pacific and Southeast).
2. Only for eligible patients between 2 and 16 years of age
3. Only for patients ≥ 14 and < 17 years of age

Aggregated Values for Q4 2020 QI Measure 4 (Immunization Status)[1]

	PCV*			PPSV*			MenACYW*			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	1531	1740	88.0	1386	1594	87.0	1272	1657	76.8	1546	181	83.5	1038	1907	54.4	841	1057	79.6	559	1510	37.0
Adult	469	660	71.1	426	666	64.0	346	577	60.0	459	750	61.2	412	756	54.5	251	540	46.5	98	535	18.3

Note: N = Numerator; D= Denominator.

1. Heartland/Southwest, Midwest, and Pacific RCCs included in aggregate for Q4 2020 Measure 4 Immunization Status. The SE region does not collect immunizations (adult or pediatric). NE provides data on a 6-month schedule.

*Vaccine priority for this project

Aggregation Across Quarters: Q3 2020 – Q4 2020

The aggregated data more thoroughly represents a population level view of the data across the nation. In order to compare across RCCs, NE data for Q2 2019 (Combined Q1/Q2: January 1, 2019 – June 30, 2019), Q4 2019 (Combined Q3/Q4: July 1, 2019 – December 31, 2019), and Q2 2020 (Combined Q1/Q2: January 1, 2020 – June 30, 2020) are included for each aggregation.

On page 6, you will find the most recent (Q3/Q4 2020) data submission in cross quarter aggregation. For a visual representation of the aggregation across quarters, see graphs starting on page 9.

Aggregation Across Quarters: Q1 – Q2 2019 (January 1-June 30, 2019)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	5249	7526	69.7	586	3548	16.5	3348	5076	66.0	434	986	44.0	568
Adult	2500	4389	57.0	679	3463	19.6							

QI Measure 4 (Immunization Status)																					
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3082	3970	77.6	3111	3770	82.5	2406	3977	60.5	3125	3989	78.3	2202	3954	55.7	1819	3664	49.6	797	3693	21.6
Adult	1189	2066	57.6	1271	2129	59.7	1103	1952	56.5	1287	2304	55.9	742	1950	38.1	371	1918	19.3	419	1889	22.2

Aggregation Across Quarters: Q3 – Q4 2019 (July 1-December 31, 2019)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	7415	10436	71.1	989	7377	13.4	4139	6129	67.5	710	1860	38.2	632
Adult	3731	6142	60.7	1200	5267	22.8							

QI Measure 4 (Immunization Status)																				
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3450	4247	81.2	3507	4235	82.8	2770	4234	65.4	3213	4215	76.2	2017	4247	47.5	2394	3429	69.8	1068	3434	31.1
Adult	992	1653	60.0	1120	1711	65.5	769	1520	50.6	850	1470	57.8	662	1520	43.6	426	1100	38.7	182	1072	17.6

Aggregation Across Quarters: Q1 – Q2 2020 (January 1-June 30, 2020)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	8140	11493	70.8	966	7796	12.4	4777	7427	64.3	804	2129	37.8	1510
Adult	3560	6390	55.7	1269	5623	22.6							

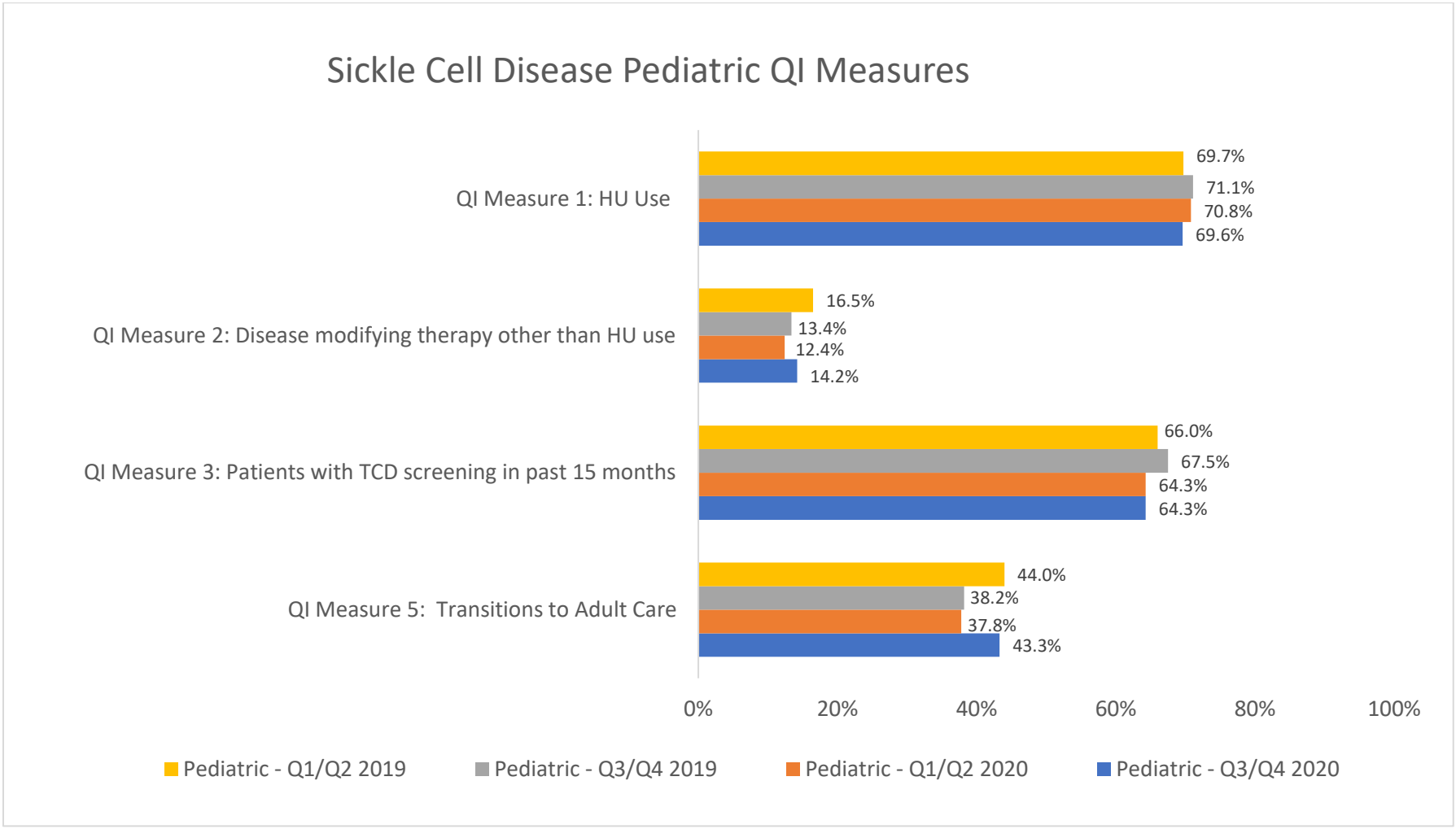
QI Measure 4 (Immunization Status)																				
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3645	4470	81.5	3367	4167	80.8	2711	4175	64.9	3323	4293	77.4	2639	4444	59.4	1798	2601	69.1	1342	3287	40.8
Adult	1428	2238	63.8	1501	2305	65.1	1263	2099	60.2	947	1570	60.3	841	1743	48.3	966	1647	58.7	429	1462	29.3

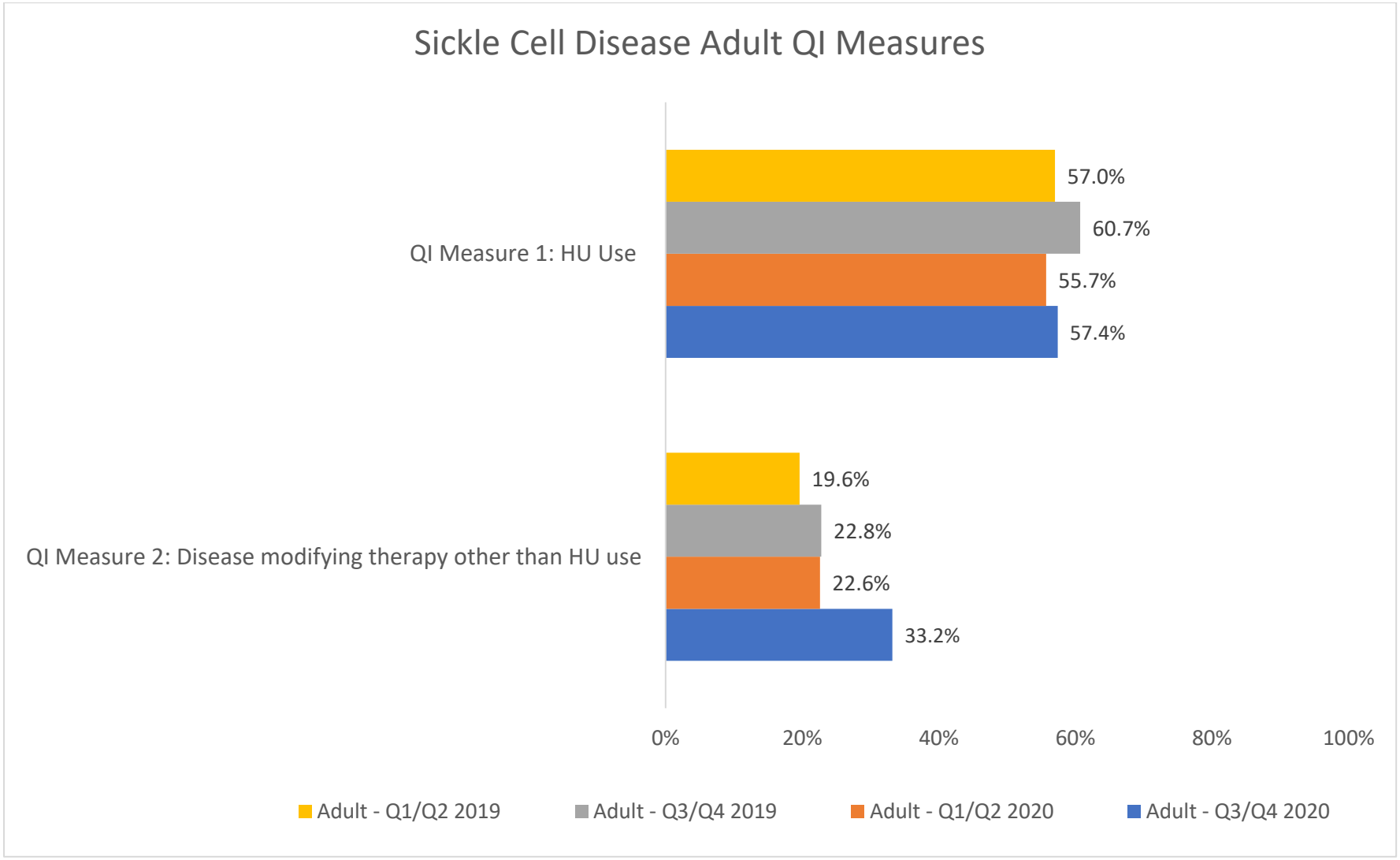
Aggregation Across Quarters: Q3 – Q4 2020 (July 1-December 31, 2020)

	QI Measure 1: HU Use			QI Measure 2: Disease modifying therapy other than HU use			QI Measure 3: Patients with TCD screening in past 15 months			QI Measure 5: Transitions to Adult Care			QI Measure 6: Providers Participating in ECHO
	N	D	%	N	D	%	N	D	%	N	D	%	Count
Pediatric	7831	11250	69.6	950	6695	14.2	4626	7195	64.3	781	1803	43.3	1035
Adult	4105	7154	57.4	1832	5518	33.2							

QI Measure 4 (Immunization Status)																				
	PCV			PPSV			MenACYW			Hib			Flu			MenB			All (PCV, PPSV, MenACYW, Hib, Flu & MenB)		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Pediatric	3326	3794	87.7	3474	4042	86.0	2770	3582	77.3	3386	4017	84.3	1948	4501	43.3	1913	2386	80.2	1129	3291	34.3
Adult	1453	2007	72.4	1400	1985	70.5	809	1496	54.1	1049	2156	48.7	798	1993	40.0	683	1418	48.2	263	1700	15.5

Graphs of Aggregation Across Quarters




Sickle Cell Disease Adult QI Measures
QI Measure 1: HU Use
57.0%
60.7%
55.7%
57.4%
QI Measure 2: Disease modifying therapy other than HU use
19.6%
22.8%
22.6%
33.2%
0%
20%
40%
60%
80%
100%
Adult - Q1/Q2 2019
Adult - Q3/Q4 2019
Adult - Q1/Q2 2020
Adult - Q3/Q4 2020

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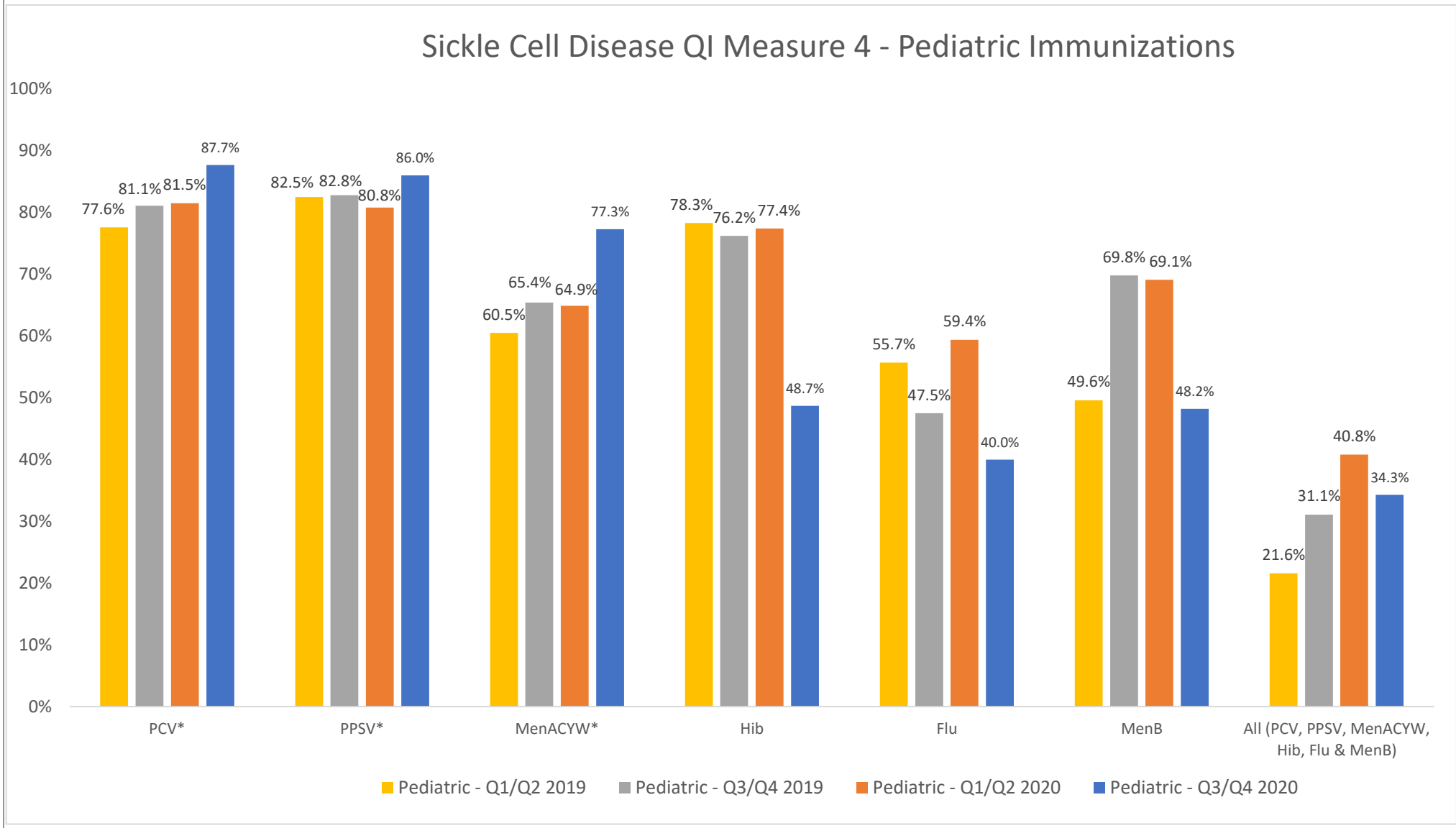
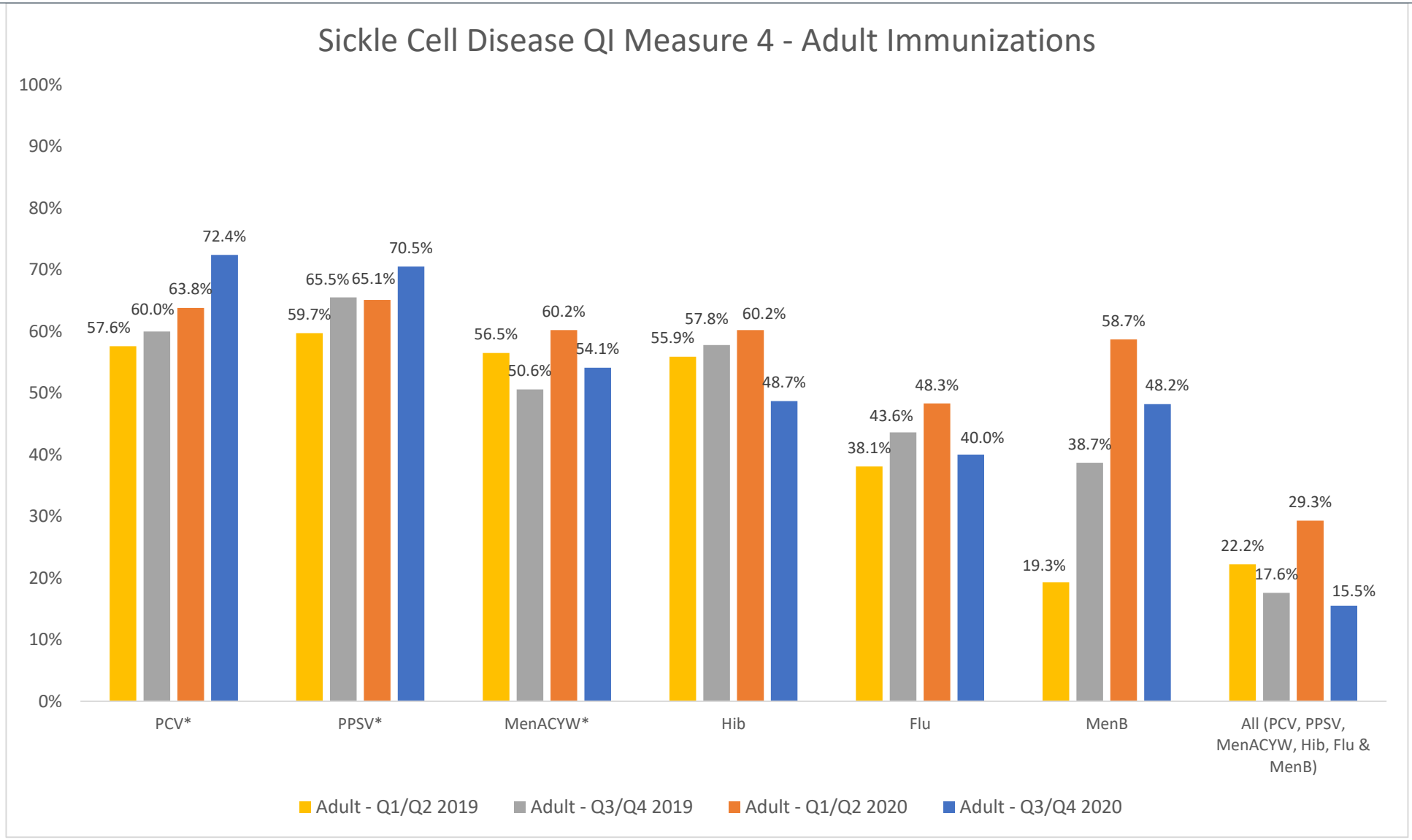
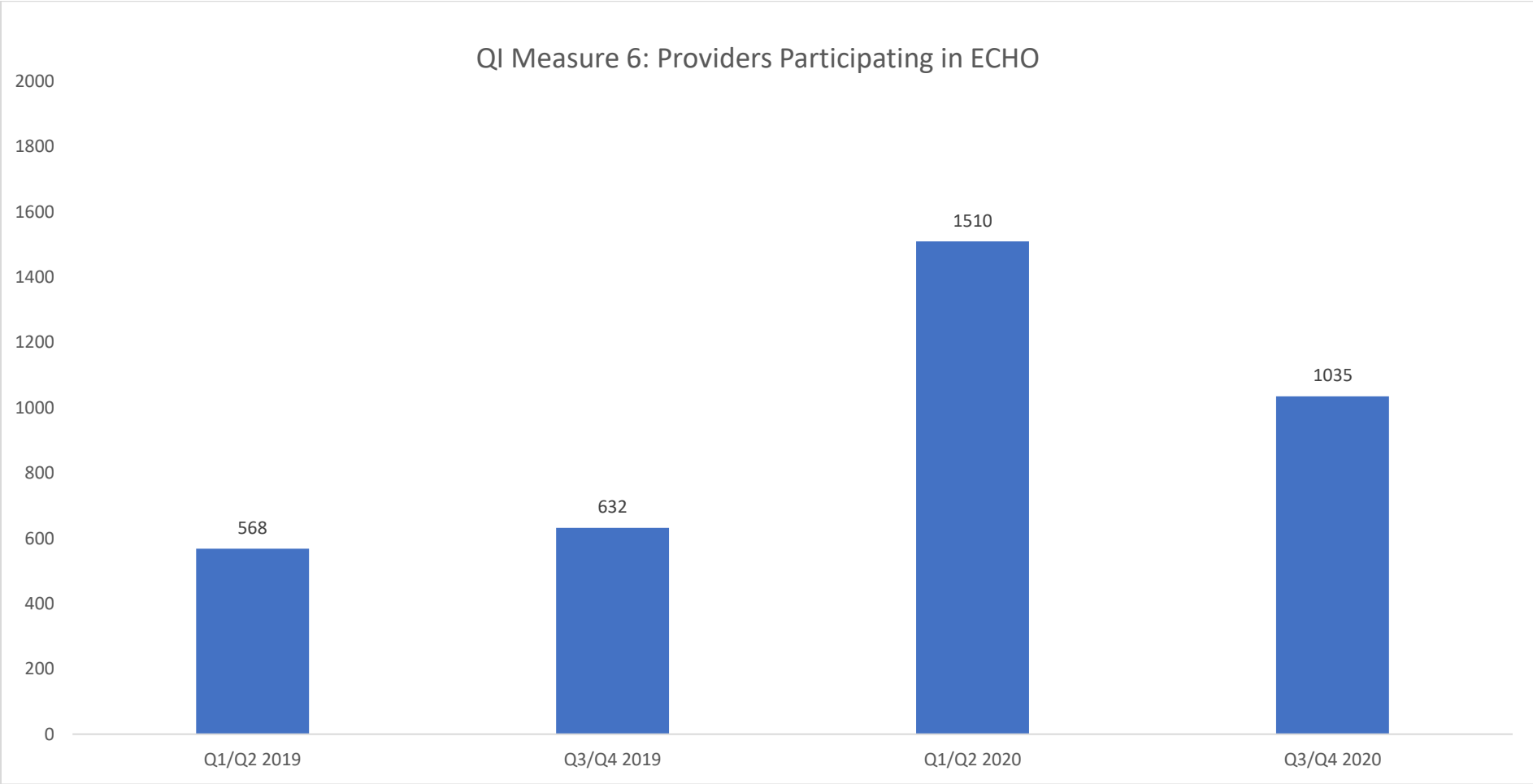

Sickle Cell Disease QI Measure 4 - Pediatric Immunizations
100%
90%
80%
70%
60%
50%
40%
30%
20%
10%
0%
77.6%
81.1%
81.5%
87.7%
82.5%
82.8%
80.8%
86.0%
60.5%
65.4%
64.9%
77.3%
78.3%
76.2%
77.4%
48.7%
55.7%
47.5%
59.4%
40.0%
49.6%
69.8%
69.1%
48.2%
21.6%
31.1%
40.8%
34.3%
PCV*
PPSV*
MenACYW*
Hib
Flu
MenB
All (PCV, PPSV, MenACYW, Hib, Flu & MenB)
Pediatric - Q1/Q2 2019
Pediatric - Q3/Q4 2019
Pediatric - Q1/Q2 2020
Pediatric - Q3/Q4 2020

11


Sickle Cell Disease QI Measure 4 - Adult Immunizations
100%
90%
80%
70%
60%
50%
40%
30%
20%
10%
0%
57.6%
60.0%
63.8%
72.4%
59.7%
65.5%
65.1%
70.5%
56.5%
50.6%
60.2%
54.1%
55.9%
57.8%
60.2%
48.7%
38.1%
43.6%
48.3%
40.0%
19.3%
38.7%
58.7%
48.2%
22.2%
17.6%
29.3%
15.5%
PCV*
PPSV*
MenACYW*
Hib
Flu
MenB
All (PCV, PPSV, MenACYW, Hib, Flu & MenB)
Adult - Q1/Q2 2019
Adult - Q3/Q4 2019
Adult - Q1/Q2 2020
Adult - Q3/Q4 2020

12


QI Measure 6: Providers Participating in ECHO
2000
1800
1600
1400
1200
1000
800
600
400
200
0
568
632
1510
1035
Q1/Q2 2019
Q3/Q4 2019
Q1/Q2 2020
Q3/Q4 2020

13

Region Specific Values for QI Measures

QI Measure 1a: Hydroxyurea use among Children/Adolescents (Required measure for all RCCs to collect)

Percent of patients >9-months and <18 years of age prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020			Q4 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	828	1144	72.4	849	1161	73.1	856	1153	74.2	904	1257	71.9	927	1258	73.7	957	1298	73.7	955	1298	73.6	937	1267	74.0
Midwest	345	437	78.9	338	393	86.0	418	537	77.8	361	439	82.2	542	665	81.5	526	645	81.6	537	661	81.2	428	532	80.5
Northeast[1]	643	883	72.8	775	1179	65.7				429	597	71.9				825	1159	71.2				678	1338	50.7
Southeast				1552	2300	67.5	1910	2734	69.9	1958	2735	71.6	1972	2877	68.5	1898	2771	68.5	1878	2669	70.4	1900	2619	72.5
Pacific	277	453	61.2	285	459	62.1	288	468	61.5	291	516	56.4	206	369	55.8	287	451	63.6	261	437	59.7	257	429	59.9

1. Note, Northeast Q1 data included September 1, 2018 – February 28th, 2019. In order to compare across all RCCs, NE data for Q2 (Combined Q1/Q2: January 1, 2019 – June 30, 2019) and Q4 (Combined Q3/Q4: July 1, 2019 – December 31, 2019) were used as proxy for the data presented for across quarters. This footnote will not be repeated for each measure, however, is applicable across all.

QI Measure 1b: Hydroxyurea use among Adults (Required measure for all RCCs to collect)

Percent of patients 18 years and older prescribed Hydroxyurea

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020			Q4 2020		
	N	D	%	N	D	%	N	N	D	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	258	426	60.6	246	446	55.2	234	428	54.7	249	468	53.2	256	469	54.6	262	473	55.4	246	438	56.2	270	466	57.9
Midwest	75	108	69.4	90	119	75.6	96	119	80.7	93	119	78.2	84	111	75.7	88	109	80.7	90	116	77.6	85	118	72.0
Northeast	485	874	55.5	437	891	49.1				541	870	62.2				746	1257	59.4				685	1557	44.0
Southeast				1157	1975	58.6	1151	1826	63.0	1156	1876	61.6	828	1605	51.6	1060	1874	56.6	1293	1843	70.2	1207	2091	57.7
Pacific	128	216	59.3	109	208	52.4	99	208	47.6	112	228	49.1	109	231	47.2	127	261	48.7	115	264	43.6	114	261	43.7

QI Measure 2a: Disease modifying therapy use other than HU among Children/Adolescents (Optional Measure)

Percent of patients ≥9 months and <18 years of age prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020			Q4 2020		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	100	402	24.9	95	388	24.5	96	374	25.7	94	413	22.8	91	408	22.3	87	393	22.1	100	401	24.9	98	397	24.7
Midwest	54	413	13.1	58	540	10.7	58	482	12.0	86	539	16.0	59	391	15.1	59	571	10.3	51	498	10.2	49	500	9.8
Northeast	62	275	22.5	51	273	18.7				26	197	13.2				39	678	5.8				34	312	10.9
Southeast				204	1402	14.6	255	2359	10.8	350	2864	12.2	303	2742	11.1	328	2613	12.6	310	2292	13.5	308	2295	13.4
Pacific	12	65	18.5	12	65	18.5	12	67	17.9	12	82	14.6												

QI Measure 2b: Disease modifying therapy use other than HU among Adults (Optional Measure)

Percent of patients 18 years and older prescribed disease modifying therapy other than HU

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020			Q4 2020		
	N	D	%	N	D	%	%	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	73	261	28.0	92	286	32.2	104	292	35.6	112	322	34.8	123	342	36.0	128	332	38.6	131	314	41.7	130	314	41.4
Midwest	21	104	20.2	12	105	11.4	13	105	12.4	20	103	19.4	18	83	21.7	12	92	13.0	12	99	12.1	24	113	21.2
Northeast	161	406	39.7	154	576	26.7				165	497	33.2				290	923	31.4				242	779	31.1
Southeast				318	2106	15.1	356	1823	19.5	424	2099	20.2	325	1785	18.2	373	2066	18.1	786	1789	43.9	507	2110	24.0
Pacific	4	12	33.3	5	13	38.5	4	16	25.0	2	10	20.0												

QI Measure 3: Transcranial Doppler (Optional Measure)

Eligible SCD patients between ages of 2-16, that had a Transcranial Doppler (TCD) screening within the 15 months prior to the end of the quarter

Region	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020			Q4 2020		
	N	D	%	N	D	%	%	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/Southwest	695	1041	66.8	722	1042	69.3	728	1031	70.6	777	1137	68.3	773	1144	67.6	769	1123	68.5	753	1155	65.2	759	1114	68.1
Midwest	299	371	80.6	389	477	81.6	393	472	83.3	415	523	79.3	461	571	80.7	451	578	78.0	445	570	78.1	468	604	77.5
Northeast	393	881	44.6	295	584	50.5				300	541	55.5				690	1463	47.2				487	973	50.1
Southeast				519	972	53.4	540	920	58.7	540	861	62.7	561	832	67.4	609	991	61.5	566	980	57.8	618	1006	61.4
Pacific	214	283	75.6	215	306	70.3	222	307	72.3	224	337	66.5	207	325	63.7	256	400	64.0	264	395	66.8	266	398	66.8

QI Measure 4a: Immunization - Percent of SCD patients <18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4a

Region	UTD[1] with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu[2]			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[3]	626	749	83.6	456	549	83.1	324	756	42.9	697	768	90.8	480	733	65.5	216	443	48.8	39	522	7.50
Midwest	355	514	69.1	402	514	78.2	398	514	77.4	351	514	68.3	346	514	67.3	460	514	89.5	225	514	43.8
Northeast	598	1493	40.1	754	1493	50.5	448	1493	30.0	525	1493	35.2	316	1493	21.2	38	1493	2.5	119	1493	8.0
Southeast																					
Pacific	18	29	62.1	17	29	58.6	20	29	69.0	6	29	20.7	16	29	55.2	13	29	44.8			

1. UTD= Up to date. This footnote will not be repeated in future tables.
2. Detailed information to match flu season with quarter were provided during Q2 2019. This footnote will not be repeated in future tables.
3. Region where estimates may not be accurate due to need to clarify how to report data if not all sites reported immunization data. Issue has since been resolved.

Quarter 2 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	761	991	76.8	817	991	82.4	528	991	53.3	819	991	82.6	596	991	60.1	494	991	49.8	101	991	10.2
Midwest	493	642	76.8	554	642	86.3	501	642	78.0	431	642	67.1	403	642	62.8	429	642	66.8	250	642	38.9
Northeast	815	1024	79.6	854	1024	83.4	620	1024	60.5	818	1024	79.9	344	1024	33.6	197	1024	19.2	182	1024	17.8
Southeast																					
Pacific	14	21	66.7	11	21	52.4	15	21	71.4	3	21	14.3	17	21	81.0	10	21	47.6			

Quarter 3 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	812	997	81.4	843	997	84.6	596	997	59.8	858	997	86.1	347	997	34.8	531	997	53.3	121	997	12.1
Midwest	534	630	84.8	494	630	78.4	501	630	79.5	428	630	67.9	361	630	57.3	487	630	77.3	216	630	34.3
Northeast																					
Southeast																					
Pacific	12	21	57.1	12	21	57.1	14	21	66.7	5	21	23.8	13	21	61.9	10	12	83.3			

Quarter 4 2019 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	919	1080	85.1	942	1080	87.2	720	1080	66.7	949	1080	87.9	518	1080	48.0	598	855	69.9	227	855	26.6
Midwest	600	661	90.8	528	656	80.5	523	652	80.2	496	661	75.0	456	661	69.0	580	648	89.5	297	661	44.9
Northeast	554	834	66.4	671	827	81.1	399	830	48.1	463	802	57.7	307	834	36.8	171	263	65.0	207	291	71.1
Southeast																					
Pacific	19	24	79.2	17	24	70.8	17	24	70.8	14	24	58.3	15	24	62.5	17	24	70.8			

Quarter 1 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	934	1102	84.8	944	1102	85.7	722	1102	65.5	970	1102	88.0	646	1102	58.6	618	876	70.5	341	876	38.9
Midwest	594	650	91.4	397	503	78.9	396	526	75.3	430	594	72.4	461	657	70.2	147	217	67.7	300	672	44.6
Northeast																					
Southeast																					
Pacific	19	21	90.5	17	21	81.0	18	21	85.7	15	21	71.4	16	21	76.2	18	21	85.7			

Quarter 2 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	926	1081	85.7	949	1081	87.8	786	1081	72.7	955	1081	88.3	646	1081	59.8	695	857	81.1	336	857	39.2
Midwest	572	617	92.7	367	461	79.6	388	510	76.1	409	560	73.0	410	603	68.0	135	200	67.5	279	630	44.3
Northeast	585	982	59.6	679	982	69.1	387	918	42.2	533	918	58.1	446	963	46.3	171	413	41.4	86	252	34.1
Southeast																					
Pacific	15	17	88.2	14	17	82.4	14	17	82.4	11	17	64.7	14	17	82.4	14	17	82.4			

Quarter 3 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	945	1091	86.6	971	1091	89.0	824	1091	75.5	970	1091	88.9	268	1091	24.6	710	861	82.5	145	861	16.8
Midwest	590	664	88.9	412	520	79.2	402	538	74.7	446	608	73.4	408	667	61.2	140	211	66.4	275	677	40.6
Northeast																					
Southeast																					
Pacific	10	14	71.4	11	14	78.6	10	14	71.4	154	182	84.6	6	14	42.9	10	14	71.4			

Quarter 4 2020 QI Measure 4a

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	926	1072	86.4	966	1072	90.1	817	1072	76.2	953	1072	88.9	526	1072	49.1	695	834	83.3	299	834	35.9
Midwest	603	663	91.0	417	517	80.7	452	580	77.9	442	606	72.9	364	662	55.0	145	218	66.5	260	676	38.5
Northeast	250	285	87.7	694	823	84.3	262	282	92.9	270	285	94.7	228	822	27.7	212	243	87.2	150	243	61.7
Southeast																					
Pacific	2	5	40.0	3	5	60.0	3	5	60.0	151	173	87.3	148	173	85.5	1	5	20.0			

QI Measure 4b: Immunization - Percent of SCD patients ≥ 18 years old who are up to date with vaccinations by vaccination series

Quarter 1 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest[1]	142	263	54.0	195	297	65.7	133	297	44.8	151	297	50.8	151	295	51.2	20	263	7.6	13	263	4.9
Midwest	104	189	55.0	129	189	68.3	139	189	73.5	96	189	50.8	115	189	60.8	131	189	69.3	58	189	30.7
Northeast	882	2009	43.9	523	2009	26.0	635	2009	31.6	658	2009	32.8	452	2009	22.5	33	2009	1.6	184	2009	9.2
Southeast																					
Pacific	11	94	11.7	35	111	31.5	10	17	58.8	142	191	74.3	10	17	58.8	7	17	41.2			

1. Region where estimates may not be accurate due to need to clarify how to report if not all sites reported immunization data. Issue has since been resolved

Quarter 2 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	168	300	56.0	199	300	66.3	150	300	50.0	159	300	53.0	154	300	51.3	34	300	11.3	24	300	8.0
Midwest	118	204	57.8	150	204	73.5	155	204	76.0	113	204	55.4	120	204	58.8	139	204	68.1	54	204	26.5
Northeast	625	933	67.0	525	933	56.3	511	933	54.8	485	933	52.0	186	933	19.9	36	933	3.9	270	933	28.9
Southeast																					
Pacific	21	83	25.3	38	95	40.0	5	12	41.7	141	190	74.2	6	12	50.0	4	12	33.3			

Quarter 3 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	184	264	69.7	200	264	75.8	153	264	58.0	159	264	60.2	122	264	46.2	59	264	22.3	31	264	11.7
Midwest	170	212	80.2	140	212	66.0	148	212	69.8	100	212	47.2	131	212	61.8	125	212	59.0	58	212	27.4
Northeast																					
Southeast																					
Pacific	23	79	29.1	39	91	42.9	4	12	33.3	125	182	68.7	6	12	50.0	3	12	25.0			

Quarter 4 2019 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	172	295	58.3	225	295	76.3	162	295	54.9	157	295	53.2	116	295	39.3	55	263	20.9	28	263	10.7
Midwest	179	213	84.0	151	213	70.9	157	213	73.7	110	213	51.6	91	213	42.7	137	213	64.3	45	213	21.1
Northeast	238	508	46.9	320	538	59.5	142	508	28.0	55	120	45.8	189	508	37.2	41	120	34.2	27	120	22.5
Southeast																					
Pacific	26	82	31.7	45	98	45.9	3	16	18.8	144	184	78.3	7	16	43.8	6	16	37.5			

Quarter 1 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	207	310	66.8	219	310	70.6	156	310	50.3	176	310	56.8	151	310	48.7	68	271	25.1	44	271	16.2
Midwest	176	214	82.2	150	214	70.1	156	214	72.9	110	214	51.4	93	214	43.5	138	214	64.5	44	214	20.6
Northeast																					
Southeast																					
Pacific	27	82	32.9	50	99	50.5	6	17	35.3	149	188	79.3	9	17	52.9	5	17	29.4			

Quarter 2 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	221	323	68.4	238	323	73.7	177	323	54.8	185	323	57.3	164	323	50.8	87	284	30.6	57	284	20.1
Midwest	194	233	83.3	154	233	66.1	155	233	66.5	107	233	49.9	92	233	39.5	152	233	65.2	42	233	18.0
Northeast	576	989	58.2	645	1026	62.9	607	989	61.4	56	125	44.8	325	633	51.3	513	615	83.4	242	460	52.6
Southeast																					
Pacific	27	87	31.0	45	100	45.0	6	13	46.2	164	177	92.7	7	13	53.8	3	13	23.1			

Quarter 3 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	239	337	70.9	254	337	75.4	197	337	58.5	202	337	59.9	71	337	21.1	102	293	34.8	35	293	12.0
Midwest	183	220	83.2	152	220	69.1	148	220	67.3	111	220	50.5	101	220	45.9	147	220	66.8	39	220	17.7
Northeast																					
Southeast																					
Pacific	28	82	34.1	45	92	48.9	8	10	80.0	145	179	81.0	5	10	50.0	5	10	50.0			

Quarter 4 2020 QI Measure 4b

Region	UTD with PCV			UTD with PPSV			UTD with MenACYW			UTD with Hib			UTD with Flu			UTD with MenB			UTD with all		
	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%	N	D	%
Heartland/ Southwest	251	356	70.5	251	356	70.5	202	356	56.7	205	356	57.6	165	356	46.3	114	319	35.7	67	319	21.0
Midwest	190	215	88.4	137	215	63.7	141	215	65.6	114	215	53.0	77	215	35.8	135	215	62.8	31	216	14.4
Northeast	534	708	75.4	523	670	78.1	110	352	31.3	132	670	19.7	209	670	31.2	178	355	50.1	91	652	14.0
Southeast																					
Pacific	28	89	31.5	38	95	40.0	3	6	50.0	140	179	78.2	170	185	91.9	2	6	33.3			

QI Measure 5: Transitions to Adult Care - Number of patients that have a documented transition education discussion

	Q1 2019			Q2 2019			Q3 2019			Q4 2019			Q1 2020			Q2 2020			Q3 2020			Q4 2020		
Region	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**	**N**	**D**	**%**
Heartland/Southwest	81	165	49.1	93	161	57.8	117	184	63.6	118	181	65.2	108	173	62.4	109	183	59.6	114	175	65.1	108	165	65.5
Midwest	9	48	18.8	21	63	33.3	16	63	25.4	18	59	30.5	20	70	28.6	15	63	23.8	19	71	26.8	17	73	23.3
Northeast	21	110	19.1	68	175	38.9				6	91	6.6				66	275	24.0				68	134	50.8
Pacific	16	23	69.6	14	22	63.6	13	19	68.4	11	18	61.1	12	17	70.6	8	13	61.5	5	9	55.6	6	10	60.0
Southeast				132	329	40.1	186	562	33.1	225	683	32.9	226	686	32.9	240	649	37.0	230	587	39.2	214	579	37.0

QI Measure 6: ECHO - Count of providers participating in project Echo meetings or Tele-mentoring calls

Region	Q1 2019	Q2 2019	Q3 2019	Q4 2019	Q1 2020	Q2 2020	Q3 2020	Q4 2020
Heartland/Southwest	61	43	58	54	35	87	64	66
Midwest	48	49	35	46	120	451	235	87
Northeast	170	242		211		580		399
Pacific	59		52	65	88	44	52	42
Southeast		66	56	55	72	33	73	17

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SCDTDRCP Performance Measures (Provider Survey) 2019

Executive Summary

Sickle cell disease (SCD) impacts 70,000 to 100,000 individuals in the United States [1] The goals of the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) are to improve 1) access to care 2) provider knowledge , and 3) coordination and service delivery (Hydroxyurea) on a national scale with the goal of improving outcomes for patients with SCD.

This report includes responses from an annual provider survey. The survey was completed by providers (516/1854= 28% response rate) who are part of five, US regional coordinating centers (RCC), who are grantees of the SCDTDRCP. After this base year (2019), this survey will be conducted for two additional years. The five grantee regions (from East Coast to West Coast) are:

- **SINERGE: the collaborative for the Northeast**, which covers: Maine, New Hampshire, Vermont, Massachusetts, Rhode Island, Connecticut, New York, New Jersey, Pennsylvania, Delaware, Maryland, Virginia, West Virginia, Puerto Rico, US Virgin Islands
- **EMBRACE: the collaborative for the Southeast**, which covers: North Carolina, South Carolina, Georgia, Florida, Alabama, Kentucky, Mississippi
- **STORM: the collaborative for the Midwest**, which covers: Ohio, Indiana, Illinois, Minnesota, Wisconsin, Michigan, North Dakota, South Dakota
- **Heartland & Southwest Sickle Cell Disease Network**, which covers: Missouri, Kansas, Iowa, Nebraska, Arkansas, Louisiana, Texas
- **Pacific Sickle Cell Regional Collaborative (PSCRC)**, which covers: California, Nevada, Arizona, Oregon, Washington, Idaho, Colorado, Wyoming, Montana

The findings in this report highlight several key elements. First, while the vast majority of SCD providers who responded serve urban regions and feel comfortable treating SCD patients, clinicians in rural settings who care for patients with SCD report feeling uncomfortable treating SCD and may need special consideration. Second, telementoring for SCD is widely considered to spread knowledge and build capacity among local provider networks by RCCs. Third, the disease altering therapy, hydroxyurea (HU), is in widespread use with over 47 percent of providers prescribing HU and 67 percent of patients with SCD being prescribed HU in the past year.

In summary, findings from this first annual SCDTDRCP Provider Survey highlight both areas of success and areas in need of improvement. The findings will help inform the work over the coming years as the five RCCs partner with their local sites and HRSA to advance the health and healthcare of patients with SCD.

[1] Hassell, K.L. Population Estimates of Sickle Cell Disease in the US. American Journal of Preventive Medicine, 2010. 38(4): p. S512-S521.

SCDTDRCP Performance Measures 2019

The following report provides information from the 2019 Performance Measures for the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP). RCCs aggregated and submitted their regional data to NICHQ, the National Coordinating Center (NCC) through CoLab. This report presents the results of the aggregated performance measures first, and then data by region following.

Provider Network Definitions:

The five Regional Coordinating Centers (RCCs) fielded an inaugural, annual provider survey. Each of the five RCCs defined their provider network independently so they could develop a survey dissemination strategy and sample that would best serve their respective region. As such, network descriptions varied by region. Regional network descriptions are included alphabetically in the table below:

Region	Network Definition
Heartland/ Southwest	The Heartland/Southwest RCC defined their network of providers as meeting one or more of the following criteria: • All providers who regularly treated SCD and prescribed HU • Were selected by the local site leads • ECHO participants, excluding contacts external to the region. The types of providers varied by clinical site. Some contacts came from specific provider list-serves (e.g., hematology/oncology providers, emergency department providers); others came from smaller lists of contacts that local sites felt would be most likely to respond. All providers were under the umbrella criteria for regularly treating SCD and prescribing HU.
Midwest	The Midwest RCC defined its network as prescribing providers within the Midwest region who met one or more of the following criteria: • Attended at least one of the region's ECHO sessions or had attended educational programs given by the state leads • Prescribing providers as selected by site leads including those that were partnering with the state leads
Northeast	The Northeast RCC defined its network as any licensed prescriber working in the region's states/territories/districts in the Northeast region. This region also included any licensed prescriber that had participated in at least one of their hosted ECHO sessions (note: these providers may have been outside of Northeast geographical region).
Pacific	The Pacific RCC defined its provider network as providers who attended their Project ECHO sessions (except residents and providers who do not prescribe HU, e.g. social workers, psychologists) as well as select others who the site leads know provide clinical care to the population with SCD in the region.

Southeast	The Southeast RCC allowed each local site/state lead to define individual strategy outreach. There were multiple ways they defined and collected their list of providers: 1) review of listservs, 2) review of state medical society membership, 3) review of rosters of local provider networks that may treat patients with SCD in emergency departments, community health centers, local hospitals and medical centers of SCD excellence, 4) Project ECHO participants, 5) outreach to known providers of SCD care in a state, 5) review of local ASH referral sites, and 6) review of contact rosters accrued from site visits, regional conferences, and local SCD meetings within the region.

The performance measures (PM) included in this survey:

PM 1: Sickle cell care providers in the RCC's Network

PM 2: Sickle cell disease patients seen by a provider in the SCDTDRCP Network in the past year

PM 3: Network providers participating in telementoring (e.g., ECHO) activities in the past year

PM 4: Sickle cell care providers in the Network that report feeling comfortable treating SCD patients

PM 5: Sickle cell care providers that prescribed Hydroxyurea (HU) in the past year

PM 6: SCD patients prescribed HU in the past year

Key trends from the aggregated data:

- PM 1: Respondents in the RCC's Network
  - Overall, 516* providers responded across all regions
  - The majority of providers (78.7 percent) are medical doctors
  - Approximately one-third (1/3) of providers only see adult or pediatric patients respectively; the remaining one-third (1/3) see both
  - Forty-three (43) percent of providers specialize in hematology or hematology oncology
  - Twelve (12) percent of providers are regional or state leads
  - The majority of providers serve urban regions (96.3 percent)
  - Most providers are either located in practices affiliated with a university or medical school (49.6 percent) or a practice affiliated with a non-profit hospital or hospital system (36.4 percent)
- PM 2: Patients seen by a provider in the SCDTDRCP Network in the past year
  - Overall, 27,078 patients with SCD were seen over the past year
  - The majority of patients (59.7 percent) were pediatric patients
  - The majority of patients were seen by either a Hematologist (40.2 percent) or a Hematologist/Oncologist (40.9 percent)

- PM 3: Respondents participating in telementoring (e.g., ECHO) activities in the past year
  - Twenty-three (22.5) percent of providers participated in telementoring for SCD in the past year
  - The majority of participating providers were medical doctors (79.3 percent)
  - Most participating providers were either Hematologists (39.7 percent) or Hematologist/Oncologists (37.9 percent)
  - Most participating providers were serving urban regions (95.7 percent)
  - Thirty-two percent of participating providers were regional or state leads
- PM 4: Providers in the Network that report feeling comfortable treating SCD patients
  - Fifty-eight (57.6) percent of providers reported feeling comfortable treating SCD patients
  - The majority of providers across provider types (excluding providers categorized as an "other" type) reported feeling comfortable treating SCD patients (55 percent to 61.1 percent)
  - Most hematologists and hematologist/oncologists report feeling comfortable treating SCD patients (85.6 percent and 84.1 percent respectively); other providers' reporting their comfort level range from 20 percent (primary care-med/peds) to 64.3 percent (primary care-emergency medicine)
  - The majority of urban serving providers reported feeling comfortable treating SCD patients (58.4 percent); while the majority of rural serving providers report not feeling comfortable treating SCD patients (56.3 percent)
- PM 5: Respondents that prescribed HU in the past year
  - Forty-eight (47.7) percent of respondents prescribed HU in the past year
  - The majority of medical doctors and nurse practitioners prescribed HU in the past year (50.5 percent and 55 percent respectively)
  - The majority of providers who specialize in hematology or hematology/oncology prescribed HU in the past year (78.9 and 82.6 percent respectively). The percentages for other provider specialties prescribing HU ranged from 4.76 percent (emergency medicine) to 45.2 percent (other providers)
  - Fifty (50) percent of rural serving providers prescribed HU in the past year, while 48.3 percent of urban serving providers prescribed HU
- PM 6: Patients prescribed HU in the past year
  - Sixty-seven (67.2) percent of SCD patients seen in the past year had a HU prescription
  - The majority of adult and pediatric patients seen had a HU prescription (57.9 percent and 73.4 percent respectively)
  - The majority of patients seen by a provider who specializes in hematology or hematology/oncology had a HU prescription (77.4 percent and 70.6 percent respectively)

* The total number of providers who responded to the survey was 516. Data presented includes all responses provided in the survey. There were some missing responses, however these were not typically annotated and thus cannot be confirmed.

Contents

Tracking Data

As noted, each of the five Regional Coordinating Centers defined their provider network independently (documented starting on page 2), which allowed them to implement a dissemination strategy and pursue a sample that best served their region. This report includes data from a convenience sample of 516 providers from the five regions. These providers saw over 27,000 patients with SCD.

Of note:

- Overall, 1,854 providers were sent the SCDTDRCP Provider Survey.
- Total of 516 providers across all regions responded to the provider survey (27.8% overall response rate).
- Regionally, the response rate was as follows:
    - Heartland/Southwest: 31.3% (51/163)
    - Midwest: 36.1% (79/219)
    - Northeast: 11.6% (34/292)
    - Pacific: 48.1% (38/79)
    - Southeast: 29.0% (314/1101)

Domain: Access to Care

Performance Measure 1a: (Total) Number of Providers in the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) Network

As 516 total providers responded to the provider survey, this number is the denominator in all PM 1 calculations below.

Provider Type	Total N	Percentage
Medical Doctor	406	78.7%
Nurse Practitioner	71	13.8%
Physician Assistant	15	2.91%
Other Provider[1]	5	0.97%

[1] Only Midwest annotated what type of "other provider." They state Certified Nurse Midwife. Pacific reported one "other provider" and Midwest noted four "other providers."

Performance Measure 1b: (Population Served) Number of Providers in the SCDTDRCP Network by population served

Population	Total N	Percentage
Adult (≥18 years of age)	178	34.5%
Pediatric (<18 years of age)	146	28.3%
Both Adult and Pediatric	170	32.9%

Performance Measure 1c: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP Network

Provider Specialty/Subspecialty	Total N	Percentage
Hematology	90	17.4%
Hematology/Oncology	132	25.6%
Primary Care-Pediatrics	56	10.9%
Primary Care- Internal Medicine	15	2.91%
Primary Care- Family Medicine	27	5.23%
Primary Care- Med/Peds	10	1.94%
Hospitalist	92	17.8%
Emergency Medicine	42	8.14%
Other Specialty	31	6.01%

Note: Examples of other specialties include: bone marrow transplant, pediatric pulmonary, ophthalmology, women's health, pulmonary/critical care, pediatric infectious diseases

Performance Measure 1d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network

Sixty-two providers responded that they were regional or state leads (out of 516 total respondents; 12 percent)

Performance Measure 1e: (Provider Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRCP Network

Location	Total N	Percentage
Rural	16	3.10%
Urban	497	96.3%

Performance Measure 1f: (Primary Practice Location-Practice Type) Number of Providers in the SCDTDRCP Network

Practice Type	Total N	Percentage
Solo Private Practice	12	2.33%
Group Private Practice	28	5.43%
Practice affiliated with a university or medical school	256	49.6%
Practice affiliated with a non-profit hospital or hospital system	188	36.4%
Practice affiliated with a for-profit hospital or hospital system	21	4.07%
Practice/clinic owned by a health maintenance organization or insurance company	3	0.58%
Federally qualified health center or community health center	10	1.94%
State or local government clinic	1	0.19%
Other	10	1.94%

Performance Measures 2: (Total) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year

Overall, providers reported seeing 27,078 sickle cell patients in the past year. Therefore, this is the denominator for all PM 2 calculations below:

Performance Measure 2a: (Age) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year

Patient Age	Total N	Percentage
Pediatric (<18 years of age)	16,166	59.7%
Adult (≥18 years of age)	10,912	40.3%

Performance Measure 2b: (Provider Specialty/Subspecialty) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year

Provider Specialty/Subspecialty	Total N	Percentage
Hematology	10,885	40.2%
Hematology/Oncology	11,082	40.9%
Primary Care-Pediatrics	457	1.69%
Primary Care- Internal Medicine	96	0.35%
Primary Care- Family Medicine	407	1.50%
Primary Care- Med/Peds	102	0.38%
Hospitalist	1,203	4.44%
Emergency Medicine	1,180	4.36%
Other	1,650	6.09%

Domain: Provider Knowledge

Performance Measures 3: (Total) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

116 providers (22.5%) responded that they participated in telementoring for SCD in the past year. Therefore, this number is used as the denominator in all PM 3 calculations below:

Performance Measure 3a: (Provider Type) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Provider Type	Total N	Percentage
Medical Doctor	92	79.3%
Nurse Practitioner	21	18.1%
Physician Assistant	2	1.72%
Other Provider	1	0.86%

Performance Measure 3b: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Provider Specialty/Subspecialty	Total N	Percentage
Hematology	46	39.7%
Hematology/Oncology	44	37.9%
Primary Care-Pediatrics	4	3.45%
Primary Care-Internal Medicine	3	2.59%
Primary Care-Family Medicine	6	5.17%
Primary Care-Med/Peds	0	0%
Hospitalist	4	3.45%
Emergency Medicine	1	0.86%
Other	8	6.90%

Performance Measure 3c: (Primary Practice Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Location	Total N	Percentage
Rural	4	3.45%
Urban	111	95.7%

Performance Measure 3d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Thirty-seven (37) providers noted that they were regional, or state leads and participated in telementoring for Sickle Cell Disease in the past year (31.9%)

Performance Measures 4: (Total) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients

Two hundred and ninety-seven (297) total providers (out of 516 total providers) responded that they felt comfortable treating patients with SCD (57.6 percent). The calculations below break down the total number of providers in each category who reported feeling comfortable treating patients with SCD.

Performance Measure 4a: (Provider Type) Percent of Providers in the SCDTDRCP Network that Report feeling Comfortable Treating Sickle Cell Patients

Provider Type	Numerator	Denominator	Percentage
Medical Doctor	248	406	61.1%
Nurse Practitioner	39	71	54.9%
Physician Assistant	9	15	60.0%
Other Provider	1	5	20.0%

Note: The numerator is the number of providers reporting being comfortable. The denominator is the total N of specified provider type.

Performance Measure 4b: (Provider Specialty/Subspecialty) Percent of Providers in the SCDTDRCP Network that Report feeling Comfortable Treating Sickle Cell Patients

Provider Specialty/Subspecialty	Numerator	Denominator	Percentage
Hematology	77	90	85.6%
Hematology/Oncology	111	132	84.1%
Primary Care – Pediatrics	14	56	25.0%
Primary Care – Internal Medicine	4	15	26.7%
Primary Care – Family Medicine	10	27	37.0%
Primary Care – Med/Peds	2	10	20.0%
Primary Care – Hospitalist	33	92	35.9%
Primary Care – Emergency Medicine	27	42	64.3%
Other	18	31	58.1%

Note: The numerator is the number of providers reporting being comfortable. The denominator is the total N of specialty type.

Performance Measure 4c: (Practice Location- Rural/Urban Providers- Zip codes) Percent of Providers in the SCDTDRCP Network that Report Feeling Comfortable Treating Sickle Cell Patients

Practice Location	Numerator	Denominator	Percentage
Rural	7	16	43.8%
Urban	293	497	58.9%

Note: The numerator is the number of respondents reporting feeling comfortable. The denominator is the total N of respondents in practice location.

Domain: Coordination and Delivery of Services (Hydroxyurea)

Performance Measures 5: (Total) Number of SCDTDRCP providers that saw at least one sickle cell patient in the past year, that prescribed hydroxyurea

Overall, regions reported that 246 (47.7%) providers saw a sickle cell patient in the past year and prescribed hydroxyurea.

Performance Measure 5a: (Provider Type) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient

Provider Type	Numerator	Denominator	Percentage
Medical Doctor	205	406	50.5%
Nurse Practitioner	39	71	54.9%
Physician Assistant	5	15	33.3%
Other Provider	0	5	0%

Note: The numerator is the number of respondents reporting that they saw at least one sickle cell patient in the last year and prescribed HU. The denominator is the total N of specified provider type.

Performance Measure 5b: (Provider Specialty/Subspecialty) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient

Provider Specialty/Subspecialty	Numerator	Denominator	Percentage
Hematology	71	90	78.9%
Hematology/Oncology	109	132	82.6%
Primary Care – Pediatrics	8	56	14.3%
Primary Care – Internal Medicine	4	15	26.7%
Primary Care – Family Medicine	11	27	40.7%
Primary Care – Med/Peds	1	10	10.0%
Primary Care – Hospitalist	27	92	29.3%
Primary Care – Emergency Medicine	2	42	4.76%
Other	14	31	45.2%

Note: The numerator is the number of respondents reporting that they saw at least one sickle cell patient in the last year and prescribed HU. The denominator is the total N of specialty type.

Performance Measure 5c: (Provider Location - Rural/Urban Zip Code Providers) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient

Practice Location	Numerator	Denominator	Percentage
Rural	8	16	50.0%
Urban	239	497	48.1%

The numerator is the number of respondents reporting that they saw at least one sickle cell patient in the last year and prescribed HU. The denominator is the total N of respondents in practice location.

Performance Measures 6: (Total) Number of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year

A total of 18,194 sickle cell patients seen by providers were noted to have a hydroxyurea prescription in the past year (67.2% of the 27,078 SCD patients seen by providers in the past year).

Performance Measure 6a: (Age) Percent of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year

Population	Numerator	Denominator	Percentage
Adult (≥18 years of age)	6,323	10,912	57.9%
Pediatric (<18 years of age)	11,871	16,166	73.4%
Total	18,194	27,078	67.2%

Note: The numerator is the number of respondents reporting population type who had HU prescription in past year. The denominator is the total N of SCD patients seen by providers in the past year.

Performance Measure 6b: (Provider Specialty/Subspecialty) Percent of sickle cell patients seen by a SCDTDRCP Network provider that had a hydroxyurea prescription in the past year

Provider Specialty/Subspecialty	Numerator	Denominator	Percentage
Hematology	8,427	10,885	77.4%
Hematology/Oncology	7,821	11,082	70.6%
Primary Care – Pediatrics	112	457	24.5%
Primary Care – Internal Medicine	52	96	54.2%
Primary Care – Family Medicine	186	407	45.7%
Primary Care – Med/Peds	10	102	9.80%
Primary Care – Hospitalist	358	1,203	29.8%
Primary Care – Emergency Medicine	2	1,180	0.17%
Other	973	1,650	59%

Note: The numerator is the number of SCD patients who had HU prescription in past year in each specialty type. The denominator is the total N of SCD patients seen by specialty type in the past year.

Regional Data

Domain: Access to Care

Performance Measures 1: (Total) Number of Providers in the SCDTDRCP Network

Region	Whole Network	Total Respondents
Southeast	1101	314
Heartland/Southwest	163	51
Northeast	292	34
Pacific	79	38
Midwest	219	79

The denominators for each of the below measures are the total respondents by region as depicted above.

Performance Measure 1a: (Population served) Number of Providers in the SCDTDRCP Network by provider type and population served

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	239/314 (76.1%)	44/51 (86.3%)	31/34 (91.2%)	28/38 (73.7%)	64/79 (81.0%)
Nurse Practitioner	41/314 (13.1%)	4/51 (7.84%)	3/34 (8.82%)	9/38 (23.7%)	14/79 (17.7%)
Physician Assistant	11/314 (3.50%)	3/51 (5.88%)	0/34 (0%)	1/38 (2.63%)	0/79 (0%)
Other Providers	4/314 (1.27%)	0/51 (0%)	0/34 (0%)	0/38 (0%)	1/79 (1.27%)

Note: N stands for numerator, D stands for denominator. Denominator is total number of providers who responded to survey per region.

Performance Measure 1b: (Population served) Number of Providers in the SCDTDRCP Network by age group seen and region served

Population	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Adult (≥18 years of age)	133/314 (42.4%)	9/51 (17.6%)	12/34 (35.3%)	16/38 (42.1%)	8/79 (10.1%)
Pediatric (<18 years of age)	75/314 (23.9%)	21/51 (41.2%)	6/34 (17.6%)	9/38 (23.7%)	35/79 (44.3%)
Both Adult and Pediatric	86/314 (27.4%)	21/51 (41.2%)	15/34 (44.1%)	13/38 (34.2%)	35/79 (44.3%)

Performance Measure 1c: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP network by specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	40/314 (12.7%)	11/51 (21.6%)	6/34 (17.6%)	14/38 (36.8%)	19/79 (24.1%)
Hematology/Oncology	51/314 (16.2%)	27/51 (52.9%)	15/34 (44.1%)	16/38 (42.1%)	23/79 (29.1%)
Primary Care – Pediatrics	35/314 (11.2%)	2/51 (3.92%)	0/34 (0%)	1/38 (2.63%)	18/79 (22.8%)
Primary Care – Internal Medicine	12/314 (3.82%)	0/51 (0%)	1/34 (2.94%)	0/38 (0%)	2/79 (2.53%)
Primary Care – Family Medicine	19/314 (6.05%)	1/51 (1.96%)	2/34 (5.88%)	2/38 (5.26%)	3/79 (3.80%)
Primary Care – Med/Peds	3/314 (0.96%)	0/51 (0%)	1/34 (2.94%)	0/38 (0%)	6/79 (7.60%)
Hospitalist	87/314 (27.7%)	0/51 (0%)	0/34 (0%)	4/38 (10.5%)	1/79 (1.27%)
Emergency Medicine	29/314 (9.24%)	6/51 (11.8%)	6/34 (17.6%)	0/38 (0%)	1/79 (1.27%)
Other	19/314 (6.05%)	4/51 (7.84%)	3/34 (8.82%)	1/38 (2.63%)	4/79 (5.06%)

Performance Measure 1d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network by Region

Region	Numerator	Denominator	Percentage
Southeast	26	314	8.28%
Heartland/Southwest	11	51	21.6%
Northeast	5	34	14.7%
Pacific	13	38	34.2%
Midwest	7	79	8.86%

Performance Measure 1e: (Provider Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRCP Network by Location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	7/314 (2.23%)	0/51 (0%)	0/34 (0%)	3/38 (7.89%)	6/79 (7.59%)
Urban	307/314 (97.8%)	51/51 (100%)	34/34 (100%)	35/38 (92.1%)	70/79 (88.6%)

Performance Measure 1f: (Primary Practice Location-Practice Type) Number of Providers in the Sickle Cell Disease Treatment SCDTDRCP Network by Practice Type

Practice Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Solo Private Practice	8/314 (2.55%)	2/51 (3.92%)	1/34 (2.94%)	0/38 (0%)	1/79 (1.27%)
Group Private Practice	15/314 (4.78%)	0/51 (0%)	4/34 (11.8%)	1/38 (2.63%)	8/79 (10.1%)
Practice affiliated with a university or medical school	144/314 (45.9%)	42/51 (82.4%)	16/34 (47.1%)	24/38 (63.2%)	30/79 (38.0%)
Practice affiliated with a non-profit hospital or hospital system	104/314 (33.1%)	14/51 (27.5%)	16/34 (47.1%)	14/38 (36.8%)	40/79 (50.6%)
Practice affiliated with a for-profit hospital or hospital system	13/314 (4.14%)	0/51 (0%)	1/34 (2.94%)	2/38 (5.26%)	5/79 (6.33%)
Practice/clinic owned by a health maintenance organization or insurance company	0/314 (0%)	0/51 (0%)	0/34 (0%)	3/38 (7.89%)	0/79 (0%)
Federally qualified health center or community health center	2/314 (0.64%)	3/51 (5.88%)	0/34 (0%)	1/38 (2.63%)	4/79 (5.06%)
State or Local Government Clinic	1/314 (0.32%)	0/51 (0%)	0/34 (0%)	0/38 (0%)	0/79 (0%)
Other	7/314 (2.23%)	0/51 (0%)	0/34 (0%)	0/38 (0%)	3/79 (3.80%)

Performance Measures 2: Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year by Region

Region	count
Southeast	15,330
Heartland/Southwest	4,426
Northeast	2,318
Pacific	1,989
Midwest	3,015

Performance Measure 2a: (Age) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year by Age Group

Population	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Adult (≥18 years of age)	6764/15,330 (44.1%)	1,594/4,426 (36.0%)	1,085/2,318 (46.8%)	938/1,989 (47.2%)	531/3,015 (17.6%)
Pediatric (<18 years of age)	8,566/15,330 (55.9%)	2,832/4,426 (64.0%)	1,233/2,318 (53.2%)	1,051/1,989 (52.8%)	2,484/3015 (82.4%)

Performance Measure 2b: (Provider Specialty/Subspecialty) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Specialty Provider in the Past Year

Provider Specialty/Subspecialty	**Southeast N/D (%)**	**Heartland/Southwest N/D (%)**	**Northeast N/D (%)**	**Pacific N/D (%)**	**Midwest N/D (%)**
Hematology	6,913/15,330 (45.1%)	1,298/4,426 (29.3%)	665/2,318 (28.7%)	597/1,989 (30.0%)	1,412/3,015 (46.8%)
Hematology/Oncology	5,026/15,330 (32.8%)	2,540/4,426 (57.4%)	1,218/2,318 (52.6%)	889/1,989 (44.7%)	1,409/3,015 (46.7%)
Primary Care – Pediatrics	249/15,330 (1.62%)	7/4,426 (0.16%)	0/2,318 (0%)	150/1,989 (7.54%)	51/3,015 (1.69%)
Primary Care – Internal Medicine	47/15,330 (0.31%)	0/4,426 (0%)	0/2,318 (0%)	0/1,989 (0%)	49/3,015 (1.63%)
Primary Care – Family Medicine	121/15,330 (0.79%)	103/4,426 (2.33%)	10/2,318 (0.43%)	166/1,989 (8.35%)	7/3,015 (0.23%)
Primary Care – Med/Peds	61/15,330 (0.40%)	0/4,426 (0%)	28/2,318 (1.21%)	0/1,989 (0%)	13/3,015 (0.43%)
Hospitalist	1,183/15,330 (7.72%)	0/4,426 (0%)	0/2,318 (0%)	20/1,989 (1.01%)	0/3,015 (0%)
Emergency Medicine	720/15,330 (4.70%)	383/4,426 (8.65%)	64/2,318 (2.76%)	0/1,989 (0%)	13/3,015 (0.43%)
Other	1,010/15,330 (6.59%)	95/4,426 (2.15%)	333/2,318 (14.4%)	167/1,989 (8.40%)	45/3,015 (1.50%)

Domain: Provider Knowledge

Performance Measures 3: (Total) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Region

Region	Count
Southeast	38
Heartland/Southwest	23
Northeast	10
Pacific	29
Midwest	16

The denominators for each of the below measures are the total respondents by region as depicted above.

Performance Measure 3a: (Provider Type) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Provider Type

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	27/38 (71.1%)	19/23 (82.6%)	10/10 (100%)	21/29 (72.4%)	15/16 (93.8%)
Nurse Practitioner	8/38 (21.1%)	4/23 (17.4%)	0/10 (0%)	8/29 (27.6%)	1/16 (6.25%)
Physician Assistant	2/38 (5.26%)	0/23 (0%)	0/10 (0%)	0/29 (0%)	0/16 (0%)
Other Providers	1/38 (2.63%)	0/23 (0%)	0/10 (0%)	0/29 (0%)	0/16 (0%)

Performance Measure 3b: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	17/38 (44.7%)	6/23 (26.1%)	2/10 (20.0%)	12/29 (41.4%)	9/16 (56.3%)
Hematology/Oncology	9/38 (23.7%)	16/23 (69.6%)	4/10 (40.0%)	10/29 (34.5%)	5/16 (31.3%)
Primary Care – Pediatrics	1/38 (2.63%)	0/23 (0%)	0/10 (0%)	1/29 (3.45%)	2/16 (12.5%)
Primary Care – Internal Medicine	3/38 (7.89%)	0/23 (0%)	0/10(0%)	0/29 (0%)	0/16 (0%)
Primary Care – Family Medicine	2/38 (5.26%)	1/23 (4.35%)	1/10 (10.0%)	2/29 (6.90%)	0/16 (0%)
Primary Care – Med/Peds	0/38 (0%)	0/23 (0%)	0/10 (0%)	0/29 (0%)	0/16 (0%)
Hospitalist	1/38 (2.63%)	0/23 (0%)	0/10 (0%)	3/29 (10.3%)	0/16 (0%)
Emergency Medicine	0/38 (0%)	0/23 (0%)	1/10 (10%)	0/29 (0%)	0/16 (0%)
Other	5/38 (13.2%)	0/23 (0%)	2/10 (20.0%)	1/29 (3.45%)	0/16 (0%)

Performance Measure 3c: (Primary Practice Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	1/38 (2.63%)	0/23 (0%)	0/10 (0%)	2/29 (6.90%)	1/16 (6.25%)
Urban	37/38 (97.4%)	23/23 (100%)	10/10 (100%)	27/29 (93.1%)	14/16 (87.5%)

Performance Measure 3d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network Who Participated in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Region	Numerator	Denominator	Percentage
Southeast	10	38	26.3%
Heartland/Southwest	7	23	30.4%
Northeast	2	10	20.0%
Pacific	12	29	41.4%
Midwest	6	16	37.5%

Performance Measures 4: (Total) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Region

Region	Count
Southeast	145
Heartland/Southwest	47
Northeast	29
Pacific	32
Midwest	44

Performance Measure 4a: (Provider Type) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Provider Type

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	120/239 (50.2%)	40/44 (90.9%)	26/31 (83.9%)	23/28 (82.1%)	39/64 (60.9%)
Nurse Practitioner	19/41 (46.3%)	4/4 (100%)	3/3 (100%)	8/9 (88.9%)	5/14 (35.7%)
Physician Assistant	5/11 (45.5%)	3/3 (100%)	0/0 (0%)	1/1 (100%)	0/0 (0%)
Other Providers	1/4 (25.0%)	0/0 (0%)	0/0 (0%)	0/0 (0%)	0/1 (0%)

Note: The numerator is the number of providers by provider type in each region who report feeling comfortable treating sickle cell patients. The denominator is the total number of provider type by region

Performance Measure 4b: (Provider Specialty/Subspecialty) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	31/40 (77.5%)	11/11 (100%)	6/6 (100%)	14/14 (100%)	15/19 (78.9%)
Hematology/Oncology	38/51 (74.5%)	26/27 (96.3%)	14/15 (93.3%)	13/16 (81.3%)	20/23 (87.0%)
Primary Care – Pediatrics	10/35 (28.6%)	2/2 (100%)	0/0 (0%)	1/1 (100%)	1/18 (5.56%)
Primary Care – Internal Medicine	3/12 (25.0%)	0/0 (0%)	0/1 (0%)	0/0 (0%)	1/2 (50.0%)
Primary Care – Family Medicine	7/19 (36.8%)	1/1 (100%)	0/2 (0%)	1/2 (50.0%)	1/3 (33.3%)
Primary Care – Med/Peds	0/3 (0%)	0/0 (0%)	1/1 (100%)	0/0 (0%)	1/6 (16.7%)
Hospitalist	31/87 (35.6%)	0/0 (0%)	0/0 (0%)	2/4 (50.0%)	0/1 (0%)
Emergency Medicine	16/29 (55.2%)	5/6 (83.3%)	5/6 (83.3%)	0/0 (0%)	1/1 (100%)
Other	9/19 (47.4%)	2/4 (50.0%)	3/3 (100%)	1/1 (100%)	3/4 (75.0%)

Note: The numerator is the number of providers by specialty in each region who report feeling comfortable treating sickle cell patients. The denominator is the total number of provider specialty by region.

Performance Measure 4c: (Practice Location- Rural/Urban Providers- Zip codes) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	3/7 (42.9%)	0/0 (0%)	0/0 (0%)	1/3 (33.3%)	3/6 (50.0%)
Urban	142/307 (46.3%)	47/51 (92.2%)	34/34 (100%)	28/35 (80.0%)	39/70 (55.7%)

Note: The numerator is the number of providers by provider location in each region who report feeling comfortable treating sickle cell patients. The denominator is the total number of provider location by region.

Domain: Coordination and Delivery of Services (Hydroxyurea)

Performance Measures 5: (Total) Number of SCDTDRCP providers that saw at least one sickle cell patient in the past year, that prescribed hydroxyurea by Region

Region	Count
Southeast	115
Heartland	43
Northeast	22
Pacific	28
Midwest	38

Performance Measure 5a: (Provider Type) Number of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient by provider type

Provider Type	Southeast N/D (%)	Heartland N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	93/239 (38.9%)	36/44 (81.8%)	20/31 (64.5%)	21/28 (75.0%)	35/64 (54.7%)
Nurse Practitioner	21/41 (51.2%)	4/4 (100%)	3/3 (100%)	6/9 (66.7%)	5/14 (35.7%)
Physician Assistant	1/11 (9.09%)	3/3 (100%)	0/0 (0%)	1/1 (100%)	0/0 (0%)
Other Providers	0/4 (0%)	0/0 (0%)	0/0 (0%)	0/0 (0%)	0/1 (0%)

Note: The numerator is the number of provider type in the SCDTDRCP network (table above) that saw at least one sickle cell patient in the last year and prescribed HU to a sickle cell patient. The denominator is the total N of specified provider type.

Performance Measure 5b: (Provider Specialty/Subspecialty) Percent of providers in the SCDTDP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient by specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	30/40 (75.0%)	11/11 (100%)	6/6 (100%)	11/14 (78.6%)	13/19 (68.4%)
Hematology/Oncology	38/51 (74.5%)	25/27 (92.6%)	15/15 (100%)	12/16 (75.0%)	19/23 (82.6%)
Primary Care – Pediatrics	3/35 (8.57%)	2/2 (100%)	0/0 (0%)	1/1 (100%)	2/18 (11.1%)
Primary Care – Internal Medicine	3/12 (25.0%)	0/0 (0%)	0/1 (0%)	0/0 (0%)	1/2 (50.0%)
Primary Care – Family Medicine	8/19 (42.1%)	1/1 (100%)	0/2 (0%)	2/2 (100%)	0/3 (0%)
Primary Care – Med/Peds	0/3 (0%)	0/0 (0%)	1/1 (100%)	0/0 (0%)	0/6 (0%)
Hospitalist	26/87 (29.9%)	0/0 (0%)	0/0 (0%)	1/4 (25.0%)	0/1 (0%)
Emergency Medicine	0/29 (0%)	2/6 (33.3%)	0/6 (0%)	0/0 (0%)	0/1 (0%)
Other	7/19 (36.8%)	2/4 (50.0%)	1/3 (33.3%)	1/1 (100%)	3/4 (75.0%)

Note: The numerator is the number of specialty type in the SCDTDRCP network that saw at least one sickle cell patient in the last year and prescribed HU to a sickle cell patient. The denominator is the total N of specialty type.

Performance Measure 5c: (Provider Location - Rural/Urban Zip Code Providers) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient by location

Provider Location	Southeast N/D (%)	Heartland N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	4/7 (57.1%)	0/0 (0%)	0/0 (0%)	2/3 (66.7%)	2/6 (33.3%)
Urban	111/307 (36.2%)	43/51 (84.3%)	22/34 (64.7%)	27/35 (77.1%)	37/70 (52.9%)

Note: The numerator is the number of provider type in the SCDTDRCP network that saw at least one sickle cell patient in the last year and prescribed HU to a sickle cell patient. The denominator is the total N of provider location type (rural or urban).

Performance Measures 6: (Total) Number of sickle cell patients seen by a SCDTDRCP Network Provider wrote a hydroxyurea prescription in the past year by region

Region	Count
Southeast	12,538
Southwest	2,197
Northeast	1,067
Pacific	943
Midwest	1,449

Performance Measure 6a: (Age) Number of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year by population

Population	Southeast N/D (%)	Heartland N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Adult (≥18 years of age)	4,518/6,764 (66.8%)	593/1,594 (37.2%)	471/1,085 (43.4%)	394/938 (42.0%)	347/531 (65.3%)
Pediatric (<18 years of age)	8,020/8,566 (93.6%)	1,604/2,832 (56.6%)	596/1,233 (48.3%)	549/1,051 (52.2%)	1,102/2,484 (44.4%)

Note: The numerator is the number of SCD patients in population group seen by a SCDTDRCP provider in past year and received a prescription for HU in the past year. The denominator is the total N of population type.

Performance Measure 6b: (Provider Specialty/Subspecialty) Number of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year by specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	6,480/6,913 (93.7%)	652/1,298 (50.2%)	217/665 (32.6%)	275/597 (46.1%)	803/1,412 (56.9%)
Hematology/Oncology	4,722/5,026 (94.0%)	1,484/2,540 (58.4%)	584/1,218 (47.9%)	471/889 (53.0%)	560/1,409 (39.7%)
Primary Care – Pediatrics	38/249 (15.3%)	5/7 (71.4%)	0/0 (0%)	60/150 (40.0%)	9/51 (17.6%)
Primary Care – Internal Medicine	14/47 (29.8%)	0/0 (0%)	0/0 (0%)	0/0 (0%)	38/49 (77.6%)
Primary Care – Family Medicine	78/121 (64.5%)	34/103 (33.0%)	0/10 (0%)	74/166 (44.6%)	0/7 (0%)
Primary Care – Med/Peds	0/61 (0%)	0/0 (0%)	4/28 (14.3%)	0/0 (0%)	6/13 (46.2%)
Hospitalist	348/1,183 (29.4%)	0/0 (0%)	0/0 (0%)	10/20 (50.0%)	0/0 (0%)
Emergency Medicine	0/720 (0%)	2/383 (0.52%)	0/64 (0%)	0/0 (0%)	0/13 (0%)
Other	857/1,010 (84.9%)	20/95 (21.1%)	12/333 (3.60%)	53/167 (31.7%)	31/45 (68.9%)

Note: The numerator is the number of SCD patients seen by SCDTDRCP specialty provider in past year and received a prescription for HU in the past year. The denominator is the total N of specialty provider type.

Appendix A: Performance Measures (PM)/Provider Survey: Lessons Learned/Challenges

The NICHQ DARE team played a key role in aligning the provider survey with the performance measures data dictionary to ensure that the intended information was collected, aggregated and submitted to CoLab. NICHQ worked closely with the HRSA counterpart charged with creating the performance measure domains and survey. Through an iterative process of review between DARE, NICHQ Data Faculty, RCCs, and the HRSA epidemiologist, alignment was arrived at and shared with the RCCs for dissemination. The end result was a final performance measure survey, corresponding performance measure data dictionary, survey distribution and CoLab aggregated regional data entry.

There has been much to learn from this inaugural implementation of the provider survey and collection of baseline data. Below are some lessons learned and areas to refine related to the collection of performance measure data.

Lesson 1: Network Definition and Survey Fielding

Fielding of the survey went smoothly for all RCCs. All RCCs received expected response rates based on their dissemination practice. Definition of network providers, or to whom they disseminated the survey, varied widely. For this start up iteration of survey dissemination, it was best that regions defined their population. NICHQ has noted the variation in network definitions for context regarding response rate and information collected this year and going forward. HRSA and the regions may elect to standardize a network definition going forward.

Lessons 2: Challenges with Data Aggregation

As expected, there were some PM data entry issues and clarifications that arose during the data entry collection and aggregation. NICHQ discussed these with RCCs and obtained input from data faculty about how to address and resolve issues. Overarching changes and solutions/resolutions that need to be made for future survey fielding have been discussed with RCCs, data faculty and HRSA. NICHQ noted the questions/clarifications and made updates to the evergreen MOP link based on issue resolution.

Lesson 3: Annotation is key to understanding context of data collection

It is important to note regional variations to more fully understand the context of aggregated data. Providing annotations to help with interpretation of data is common in measurement, and therefore was easily accommodated by CoLab. NICHQ will continue to remind and request RCCs to heavily annotate variations, reasons for missing data and other notable details to help readers best understand those data presented.

SCDTDRCP Performance Measures (Provider Survey) 2020

Executive Summary

Sickle cell disease (SCD) impacts 70,000 to 100,000 individuals in the United States [1] The goals of the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP) are to improve 1) access to care 2) provider knowledge, and 3) coordination and service delivery (Hydroxyurea, Transcranial Doppler Screening, Immunizations and Transitions to Adult Care) on a national scale with the goal of improving outcomes for patients with SCD.

This report includes responses from the second annual provider survey which provides information about Performance Measures for this project. The survey was conducted from September 1-October 13, 2020. The initial survey was conducted in 2019. Providers from the five SCDTDRCP funded regions, which cover the US and territories and are led by a regional coordinating center (RCC), responded to the survey. The response rate was 306/1220= 25.1%.

The five grantee regions (from East Coast to West Coast) are:

- **SINERGE: the collaborative for the Northeast**, which covers: Connecticut, Delaware, Maine, Maryland, Massachusetts, New Hampshire, New Jersey, New York, Pennsylvania, Puerto Rico, Rhode Island, US Virgin Islands, Vermont, Virginia, Washington DC, West Virginia
- **EMBRACE: the collaborative for the Southeast**, which covers: Alabama, Florida, Georgia, Kentucky, Mississippi, North Carolina, South Carolina, Tennessee
- **STORM: the collaborative for the Midwest**, which covers: Illinois, Indiana, Michigan, Minnesota, North Dakota, Ohio, South Dakota, Wisconsin,
- **Heartland & Southwest Sickle Cell Disease Network**, which covers: Arkansas, Iowa, Kansas, Louisiana, Missouri, Nebraska, Oklahoma, Texas
- **Pacific Sickle Cell Regional Collaborative (PSCRC)**, which covers: Alaska, Arizona, California, Colorado, Hawaii, Idaho, New Mexico, Montana, Nevada, Oregon, Utah, Washington, Wyoming

The answers collected from this survey reflect the following findings.

- While the vast majority of SCD providers who responded serve urban regions and feel comfortable treating SCD patients, clinicians in rural settings who care for patients with SCD report feeling uncomfortable treating SCD and may need special consideration.
- Telementoring for SCD is widely used as a way to spread knowledge and build capacity among local provider networks
- There is strong support for the use of the disease altering therapy, hydroxyurea (HU). Over 65 percent of providers who are able to prescribe reported doing so and 77 percent of patients with SCD had been given a prescription for HU in the past year.

[1] Hassell, K.L. Population Estimates of Sickle Cell Disease in the US. American Journal of Preventive Medicine, 2010. 38(4): p. S512-S521.

All surveys have limitations. The following items should be noted when reading the survey findings:

1) There was a revision to the network definition which guided to whom the survey should be offered. This may have impacted the response rate.

2) Several measures have small denominators and therefore caution should be taken when interpreting.

In summary, findings from this second annual SCDTDRCP Provider Survey highlight both areas of success and areas in need of improvement. The findings may help inform the work over the coming years as the five RCCs partner with their local sites and HRSA work to advance the health and healthcare of patients living with SCD.

SCDTDRCP Performance Measures 2020

The following report provides information from the annual provider survey which reflects 2020 Performance Measures for the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program (SCDTDRCP). RCCs initiated surveys to local providers and then aggregated and submitted their regional data to the National Institute for Children's Health Quality (NICHQ), the National Coordinating Center (NCC) through CoLab. NICHQ then further aggregated regional data for a national perspective. This report presents the results of the aggregated performance measures first, and then data by region.

Provider Network Definitions:

In 2019, the five Regional Coordinating Centers (RCCs) fielded an inaugural, annual provider survey. For this survey each of the five RCCs defined their own provider network used to guide the dissemination list for the survey. RCCs then developed a survey dissemination strategy that would best serve their respective region. As such, network descriptions varied by region and it was decided that a uniform definition across regions would be agreed upon prior to the next survey launch.

For the 2020 fielding of the provider survey, HRSA developed specific guidance regarding the dissemination of the survey. For this survey, eligible participants included "SCDTDRCP Providers", defined as those providers for whom SCDTDRCP funding could conceivably lead to causing changes including whether they see SCD patients, their comfort seeing SCD patients, or prescription of disease modifying therapies, particularly hydroxyurea. In other words, providers (of all specialty) were included for whom RCC's could identify meaningful contacts with the program. Based on RCC recommended touchpoints, the "SCDTDRCP providers" include those that:

- Participated in SCDTDRCP-sponsored ECHO sessions
- SCDTDRCP state leads
- Attended SCDTDRCP CME/MOC presentations or grand rounds
- Have clinical care discussions and/or care coordination with SCDTDRCP state leads
- Participated in SCDTDRCP-sponsored intensive education (e.g., boot camp)
- Participated in SCDTDRCP-sponsored provider summit and trainings
- Participated in SCDTDRCP-led QI projects
- Attended SCDTDRCP-led resident teaching

The performance measures (PM) included in the provider survey:

PM 1: Sickle cell care providers in the RCC's Network
PM 2: Sickle cell disease patients seen by a provider in the SCDTDRCP Network in the past year
PM 3: Network providers participating in telementoring (e.g., ECHO) activities in the past year
PM 4: Sickle cell care providers in the Network that report feeling comfortable treating SCD patients
PM 5: Sickle cell care providers that prescribed Hydroxyurea (HU) in the past year
PM 6: SCD patients prescribed HU in the past year

Key trends from the aggregated data from the 5 RCCs:

- PM 1: Respondents in the RCCs' Networks
  - Overall, 306* providers responded across all regions
  - The majority of providers (77.1 percent) are medical doctors
  - Approximately one-third (1/3) of providers only see adult or pediatric patients respectively; the remaining one-third (1/3) see both
  - Seventy-one (71) percent of providers specialize in hematology or hematology/oncology
  - Twenty (19.5) percent of providers are regional or state leads
  - The majority of providers serve urban regions (89.8 percent)
  - Most providers are either located in practices affiliated with a university or medical school (63.4 percent) or a practice affiliated with a non-profit hospital or hospital system (32.4 percent)
- PM 2: Patients seen by a provider in the SCDTDRCP Network in the past year
  - Overall, 25,712 patients with SCD were seen over the past year
  - The majority of patients (52.7 percent) were adult patients
  - The majority of patients were seen by either a Hematologist (54.8 percent) or a Hematologist/Oncologist (31.4 percent)
- PM 3: Respondents participating in telementoring (e.g., ECHO) activities in the past year
  - Fifty-three (52.6) percent of providers participated in telementoring for SCD in the past year
  - The majority of participating providers were medical doctors (70.8 percent)
  - Most participating providers were either Hematologists (39.8 percent) or Hematologist/Oncologists (34.2 percent)
  - The vast majority of participating providers were serving urban regions (92.5 percent)
  - Thirty (29.8) percent of participating providers were regional or state leads
- PM 4: Providers in the Network that report feeling comfortable treating SCD patients
  - Seventy-two (72.2) percent of providers reported feeling comfortable treating SCD patients
  - The majority of providers across provider types reported feeling comfortable treating SCD patients (66.7 percent to 80.0 percent)
  - Most hematologists and hematologist/oncologists report feeling comfortable treating SCD patients (92.0 percent and 71.3 percent respectively); other providers' reporting their comfort level range from 40.0 percent (2 out of 5 primary care-med/peds providers) to 100.0 percent (7 out of 7 primary care-emergency medicine providers)
  - The majority (200 out of 277) of urban serving providers reported feeling comfortable treating SCD patients (72.2 percent), whereas 62.5 percent of rural serving providers reported feeling comfortable treating SCD patients (5 out of 8 providers).
- PM 5: Respondents that prescribed HU in the past year
  - Sixty-five (65.0) percent of respondents prescribed HU in the past year
  - The majority of medical doctors, nurse practitioners and physician assistants prescribed HU in the past year (62.7 percent, 70.2 percent and 70.0 percent respectively).
  - The majority of providers who specialize in hematology or hematology/oncology prescribed HU in the past year (80.7 and 67.4 percent respectively). The percentages for

other provider specialties prescribing HU ranged from 14.3 percent (emergency medicine) to 55.6 percent (primary care- internal medicine)
    - Sixty-three (63.2) percent of urban serving providers prescribed HU in the past year, while 37.5 percent of rural serving providers prescribed HU (3 out of 8 providers responded that they have prescribed HU in the past year).
- PM 6: Patients prescribed HU in the past year
    - Seventy-seven (77.1) percent of SCD patients seen in the past year were given a HU prescription
    - The majority of adult and pediatric patients seen had a HU prescription (74.8 percent and 79.3 percent respectively)
    - The majority of patients seen by a provider who specializes in hematology or hematology/oncology had a HU prescription (81.3 percent and 75.2 percent respectively)

* The total number of providers who responded to the survey was 306. Data presented includes all responses provided in the survey. There were some missing responses, which were not all annotated.

Contents

Tracking Data

This report includes data from a convenience sample of 306 providers from the five regions. These providers saw over 25,000 patients living with SCD.

Of note:

- Overall, 1,220 providers were sent the SCDTDRCP Provider Survey.
- Total of 306 providers across all regions responded to the provider survey (25.1% overall response rate).
- Regionally, the response rate was as follows:
  - Heartland/Southwest: 30.2% (73/241)
  - Midwest: 31.3% (50/160)
  - Northeast: 16.1% (61/378)
  - Pacific: 47.3% (43/91)
  - Southeast: 22.6% (79/350)

Domain: Access to Care

Performance Measure 1a: (Total) Number of Providers in the Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) Network

As 306 total providers responded to the provider survey, this number is the denominator in all PM 1 calculations below.

Provider Type	Total N	Percentage
Medical Doctor	236	77.1%
Nurse Practitioner	57	18.6%
Physician Assistant	10	3.3%
Other Provider[1]	3	3.8%

[1] No region annotated which providers are included as "other providers."

Performance Measure 1b: (Population Served) Number of Providers in the SCDTDRCP Network by population served

Population	Total N	Percentage
Adult (≥18 years of age)	110	35.9%
Pediatric (<18 years of age)	87	28.4%
Both Adult and Pediatric	110	35.9%

Performance Measure 1c: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP Network

Provider Specialty/Subspecialty	Total N	Percentage
Hematology	88	28.8%
Hematology/Oncology	129	42.2%
Primary Care-Pediatrics	15	4.9%
Primary Care- Internal Medicine	9	2.9%
Primary Care- Family Medicine	13	4.2%
Primary Care- Med/Peds	9	2.9%
Hospitalist	5	1.6%
Emergency Medicine	7	2.3%
Other Specialty	28	9.2%

Note: Examples of other specialties include pediatric pulmonary, pulmonary/critical care, pediatric infectious diseases, Acute Pain Management general practitioner working in sickle cell clinic

Performance Measure 1d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network

Sixty providers responded that they were regional or state leads (out of 306 total respondents; 19.6 percent)

Performance Measure 1e: (Provider Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRCP Network

Location	Total N	Percentage
Rural	8	2.6%
Urban	277	89.8%

Performance Measure 1f: (Primary Practice Location-Practice Type) Number of Providers in the SCDTDRCP Network

Practice Type	Total N	Percentage
Solo Private Practice	5	1.6%
Group Private Practice	11	3.6%
Practice affiliated with a university or medical school	194	63.4%
Practice affiliated with a non-profit hospital or hospital system	99	32.4%
Practice affiliated with a for-profit hospital or hospital system	18	5.9%
Practice/clinic owned by a health maintenance organization or insurance company	1	0.3%
Federally qualified health center or community health center	9	2.9%
State or local government clinic	6	2.0%
Other	1	0.3%

Performance Measures 2: (Total) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year

Overall, providers reported seeing 25,712 sickle cell patients in the past year. Therefore, this is the denominator for all PM 2 calculations below:

Performance Measure 2a: (Age) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year

Patient Age	Total N	Percentage
Pediatric (<18 years of age)	12,171	47.3%
Adult (≥18 years of age)	13,541	52.7%

Performance Measure 2b: (Provider Specialty/Subspecialty) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year

Provider Specialty/Subspecialty	Total N	Percentage
Hematology	14,081	54.8%
Hematology/Oncology	8,071	31.4%
Primary Care-Pediatrics	197	0.8%
Primary Care- Internal Medicine	447	1.7%
Primary Care- Family Medicine	233	0.9%
Primary Care- Med/Peds	125	0.5%
Hospitalist	560	2.2%
Emergency Medicine	262	1.0%
Other	1,681	6.5%

Domain: Provider Knowledge

Performance Measures 3: (Total) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

161 providers (52.6%) responded that they participated in telementoring for SCD in the past year. Therefore, this number is used as the denominator in all PM 3 calculations below:

Performance Measure 3a: (Provider Type) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Provider Type	Total N	Percentage
Medical Doctor	114	70.8%
Nurse Practitioner	35	21.7%
Physician Assistant	9	5.6%
Other Provider	3	1.9%

Performance Measure 3b: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Provider Specialty/Subspecialty	Total N	Percentage
Hematology	64	39.8%
Hematology/Oncology	55	34.2%
Primary Care-Pediatrics	4	2.5%
Primary Care-Internal Medicine	4	2.5%
Primary Care-Family Medicine	6	3.7%
Primary Care-Med/Peds	3	1.9%
Hospitalist	3	1.9%
Emergency Medicine	1	0.6%
Other	19	11.8%

Examples of other providers include: Pulmonary, Pediatric Pulmonary and Sleep

Performance Measure 3c: (Primary Practice Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Location	Total N	Percentage
Rural	4	2.5%
Urban	149	92.5%

Performance Measure 3d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Forty-eight (48) providers noted that they were regional, or state leads and participated in telementoring for Sickle Cell Disease in the past year (29.8%)

Performance Measures 4: (Total) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients

Two-hundred and twenty-one (221) total providers (out of 306 total providers) responded that they felt comfortable treating patients with SCD (72.2 percent). The calculations below break down the total number of providers in each category who reported feeling comfortable treating patients with SCD.

Performance Measure 4a: (Provider Type) Percent of Providers in the SCDTDRCP Network that Report feeling Comfortable Treating Sickle Cell Patients

Provider Type	Numerator	Denominator	Percentage
Medical Doctor	168	236	71.2%
Nurse Practitioner	43	57	75.4%
Physician Assistant	8	10	80.0%
Other Provider	2	3	66.7%

Note: The numerator is the number of providers reporting being comfortable. The denominator is the total N of specified provider type.

Performance Measure 4b: (Provider Specialty/Subspecialty) Percent of Providers in the SCDTDRCP Network that Report feeling Comfortable Treating Sickle Cell Patients

Provider Specialty/Subspecialty	Numerator	Denominator	Percentage
Hematology	81	88	92.0%
Hematology/Oncology	92	129	71.3%
Primary Care – Pediatrics	6	15	40.0%
Primary Care – Internal Medicine	4	9	44.4%
Primary Care – Family Medicine	7	13	53.8%
Primary Care – Med/Peds	3	9	33.3%
Primary Care – Hospitalist	2	5	40.0%
Primary Care – Emergency Medicine	7	7	100.0%
Other	18	28	64.3%

Note: The numerator is the number of providers reporting being comfortable. The denominator is the total N of specialty type.

Performance Measure 4c: (Practice Location- Rural/Urban Providers- Zip codes) Percent of Providers in the SCDTDRCP Network that Report Feeling Comfortable Treating Sickle Cell Patients

Practice Location	Numerator	Denominator	Percentage
Rural	5	8	62.5%
Urban	200	277	72.2%

Note: The numerator is the number of respondents reporting feeling comfortable. The denominator is the total N of respondents in practice location.

Domain: Coordination and Delivery of Services (Hydroxyurea)

Performance Measures 5: (Total) Number of SCDTDRCP providers that saw at least one sickle cell patient in the past year, that prescribed hydroxyurea

Overall, regions reported that 199 (65.0%) providers saw a sickle cell patient in the past year and prescribed hydroxyurea.

Performance Measure 5a: (Provider Type) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient

Provider Type	Numerator	Denominator	Percentage
Medical Doctor	148	236	62.7%
Nurse Practitioner	40	57	70.2%
Physician Assistant	7	10	70.0%
Other Provider	1	3	33.3%

Note: The numerator is the number of respondents reporting that they saw at least one sickle cell patient in the last year and prescribed HU. The denominator is the total N of specified provider type.

Performance Measure 5b: (Provider Specialty/Subspecialty) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient

Provider Specialty/Subspecialty	Numerator	Denominator	Percentage
Hematology	71	88	80.7%
Hematology/Oncology	87	129	67.4%
Primary Care – Pediatrics	6	15	40.0%
Primary Care – Internal Medicine	5	9	55.6%
Primary Care – Family Medicine	5	13	38.5%
Primary Care – Med/Peds	2	9	22.2%
Primary Care – Hospitalist	2	5	40.0%
Primary Care – Emergency Medicine	1	7	14.3%
Other	13	28	46.4%

Note: The numerator is the number of respondents reporting that they saw at least one sickle cell patient in the last year and prescribed HU. The denominator is the total N of specialty type.

Performance Measure 5c: (Provider Location - Rural/Urban Zip Code Providers) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient

Practice Location	Numerator	Denominator	Percentage
Rural	3	8	37.5%
Urban	175	277	63.2%

The numerator is the number of respondents reporting that they saw at least one sickle cell patient in the last year and prescribed HU. The denominator is the total N of respondents in practice location.

Performance Measures 6: (Total) Number of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year

A total of 19,834 sickle cell patients seen by providers were noted to have a hydroxyurea prescription in the past year (77.1% of the 25,712 SCD patients seen by providers in the past year).

Performance Measure 6a: (Age) Percent of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year

Population	Numerator	Denominator	Percentage
Adult (≥18 years of age)	9,102	12,171	74.8%
Pediatric (<18 years of age)	10,732	13,541	79.3%
Total	19,834	25,712	77.1%

Note: The numerator is the number of respondents reporting population type who had HU prescription in past year. The denominator is the total N of SCD patients seen by providers in the past year.

Performance Measure 6b: (Provider Specialty/Subspecialty) Percent of sickle cell patients seen by a SCDTDRCP Network provider that had a hydroxyurea prescription in the past year

Provider Specialty/Subspecialty	Numerator	Denominator	Percentage
Hematology	11,445	14,081	81.3%
Hematology/Oncology	6,072	8,071	75.2%
Primary Care – Pediatrics	77	197	39.1%
Primary Care – Internal Medicine	290	447	64.9%
Primary Care – Family Medicine	137	233	58.8%
Primary Care – Med/Peds	102	125	81.6%
Primary Care – Hospitalist	518	560	92.5%
Primary Care – Emergency Medicine	2	262	0.8%
Other	1,136	1,681	67.6%

Note: The numerator is the number of SCD patients who had HU prescription in past year in each specialty type. The denominator is the total N of SCD patients seen by specialty type in the past year.

Regional Data

Domain: Access to Care

Performance Measures 1: (Total) Number of Providers in the SCDTDRCP Network

Region	Whole Network	Total Respondents
Southeast	350	79
Heartland/Southwest	241	73
Northeast	378	61
Pacific	91	43
Midwest	160	50

The denominators for each of the below measures are the total respondents by region as depicted above.

Performance Measure 1a: (Population served) Number of Providers in the SCDTDRCP Network by provider type and population served

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	54/79 (68.4%)	66/73 (90.4%)	42/61 (68.9%)	32/43 (74.4%)	42/50 (84.0%)
Nurse Practitioner	19/79 (24.1%)	7/73 (9.6%)	14/61 (23.0%)	10/43 (23.3%)	7/50 (14.0%)
Physician Assistant	3/79 (3.8%)	0/73 (0.0%)	5/61 (8.2%)	1/43 (2.3%)	1/50 (2.0%)
Other Providers	3/79 (3.8%)	0/73 (0.0%)	0/61 (0%)	0/43 (0.0%)	0/50 (0.0%)

Note: N stands for numerator, D stands for denominator. Denominator is total number of providers who responded to survey per region.

Performance Measure 1b: (Population served) Number of Providers in the SCDTDRCP Network by age group seen and region served

Population	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Adult (≥18 years of age)	31/79 (39.2%)	24/73 (32.9%)	32/61 (52.5%)	16/43 (37.2%)	7/50 (14.0%)
Pediatric (<18 years of age)	16/79 (20.3%)	33/73 (45.2%)	11/61 (18.0%)	11/43 (25.6%)	16/50 (32.0%)
Both Adult and Pediatric	32/79 (40.5%)	16/73 (21.9%)	19/61 (31.1%)	16/43 (37.2%)	27/50 (54.0%)

Performance Measure 1c: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP network by specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	28/79 (35.4%)	14/73 (19.2%)	19/61 (31.1%)	15/43 (34.9%)	12/50 (24.0%)
Hematology/Oncology	24/79 (30.4%)	46/73 (63.0%)	17/61 (27.9%)	21/43 (48.8%)	21/50 (42.0%)
Primary Care – Pediatrics	3/79 (3.8%)	2/73 (2.7%)	3/61 (4.9%)	1/43 (2.3%)	6/50 (12.0%)
Primary Care – Internal Medicine	4/79 (5.1%)	1/73 (1.4%)	2/61 (3.3%)	1/43 (2.3%)	1/50 (2.0%)
Primary Care – Family Medicine	5/79 (6.3%)	2/73 (2.7%)	2/61 (3.3%)	1/43 (2.3%)	3/50 (6.0%)
Primary Care – Med/Peds	3/79 (3.8%)	1/73 (1.4%)	1/61 (1.6%)	1/43 (2.3%)	3/50 (6.0%)
Hospitalist	4/79 (5.1%)	0/73 (0.0%)	0/61 (0.0%)	1/43 (2.3%)	0/50 (0.0%)
Emergency Medicine	1/79 (1.3%)	3/73 (4.1%)	2/61 (3.3%)	0/43 (0.0%)	1/50 (2.0%)
Other	7/79 (8.9%)	4/73 (5.5%)	12/61 (19.7%)	2/43 (4.7%)	3/50 (6.0%)

Performance Measure 1d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network by Region

Region	Numerator	Denominator	Percentage
Southeast	15	79	19.0%
Heartland/Southwest	11	73	15.1%
Northeast	14	61	23.0%
Pacific	13	43	30.2%
Midwest	7	50	14.0%

Performance Measure 1e: (Provider Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRCP Network by Location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	0/79 (0.0%)	2/73 (2.7%)	0/61 (0.0%)	3/43 (7.0%)	3/50 (6.0%)
Urban	79/79 (100.0%)	71/73 (97.3%)	40/61 (65.6%)	40/43 (93.0%)	47/50 (94.0%)

Performance Measure 1f: (Primary Practice Location-Practice Type) Number of Providers in the Sickle Cell Disease Treatment SCDTDRCP Network by Practice Type

Practice Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Solo Private Practice	1/79 (1.3%)	1/73 (1.4%)	2/61 (3.3%)	0/43 (0.0%)	1/50 (2.0%)
Group Private Practice	2/79 (2.5%)	1/73 (1.4%)	1/61 (1.6%)	1/43 (2.3%)	6/50 (12.0%)
Practice affiliated with a university or medical school	40/79 (50.6%)	63/73 (86.3%)	47/61 (77.0%)	24/43 (55.8%)	20/50 (40.0%)
Practice affiliated with a non-profit hospital or hospital system	26/79 (32.9%)	17/73 (23.3%)	14/61 (23.0%)	14/43 (32.6%)	28/50 (56.0%)
Practice affiliated with a for-profit hospital or hospital system	5/79 (6.3%)	4/73 (5.5%)	5/61 (8.2%)	1/43 (2.3%)	3/50 (6.0%)
Practice/clinic owned by a health maintenance organization or insurance company	0/79 (0.0%)	0/73 (0.0%)	0/61 (0.0%)	1/43 (2.3%)	0/50 (0.0%)
Federally qualified health center or community health center	1/79 (1.3%)	1/73 (1.4%)	1/61 (1.6%)	3/43 (7.0%)	3/50 (6.0%)
State or Local Government Clinic	3/79 (3.8%)	0/73 (0.0%)	2/61 (3.3%)	1/43 (2.3%)	0/50 (0.0%)
Other	1/79 (1.3%)	0/73 (0.0%)	0/61 (0.0%)	0/43 (0.0%)	0/50 (0.0%)

Performance Measures 2: Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year by Region

Region	count
Southeast	12,265
Heartland/Southwest	4,297
Northeast	4,208
Pacific	2,257
Midwest	2,685

Performance Measure 2a: (Age) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Provider in the Past Year by Age Group

Population	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Adult (≥18 years of age)	6,603/12,265 (53.8%)	2,225/4,297 (51.8%)	3,166/4,208 (75.2%)	1,042/2,257 (46.2%)	505/2,685 (18.8%)
Pediatric (<18 years of age)	5,662/12,265 (46.2%)	2,072/4,297 (48.2%)	1,042/4,208 (24.8%)	1,215/2,257 (53.8%)	2,180/2,685 (81.2%)

Performance Measure 2b: (Provider Specialty/Subspecialty) Number of Sickle Cell Patients Seen by a SCDTDRCP Network Specialty Provider in the Past Year

Provider Specialty/Subspecialty	**Southeast N/D (%)**	**Heartland/Southwest N/D (%)**	**Northeast N/D (%)**	**Pacific N/D (%)**	**Midwest N/D (%)**
Hematology	8,133/12,265 (66.3%)	1,968/4,297 (45.8%)	1,932/4,208 (45.9%)	910/2,257 (40.3%)	1,138/2,685 (42.4%)
Hematology/Oncology	2,575/12,265 (21.0%)	1,942/4,297 (45.2%)	1,294/4,208 (30.7%)	980/2,257 (43.4%)	1,280/2,685 (47.7%)
Primary Care – Pediatrics	11/12,265 (0.1%)	8/4,297 (0.2%)	66/4,208 (1.6%)	110/2,257 (4.9%)	2/2,685 (0.1%)
Primary Care – Internal Medicine	181/12,265 (1.5%)	10/4,297 (0.2%)	1/4,208 (0.02%)	200/2,257 (8.9%)	55/2,685 (2.1%)
Primary Care – Family Medicine	19/12,265 (0.2%)	94/4,297 (2.2%)	77/4,208 (1.8%)	40/2,257 (1.8%)	3/2,685 (0.1%)
Primary Care – Med/Peds	102/12,265 (0.8%)	8/4,297 (0.2%)	5/4,208 (0.1%)	6/2,257 (0.3%)	4/2,685 (0.2%)
Hospitalist	550/12,265 (4.5%)	0/4,297 (0.0%)	0/4,208 (0.0%)	10/2,257 (0.4%)	0/2,685 (0.0%)
Emergency Medicine	60/12,265 (0.5%)	141/4,297 (3.3%)	48/4,208 (1.1%)	0/2,257 (0.0%)	13/2,685 (0.5%)
Other	634/12,265 (5.2%)	126/4,297 (2.9%)	730/4,208 (17.3%)	1/2,257 (0.04%)	190/2,685 (7.1%)

Domain: Provider Knowledge

Performance Measures 3: (Total) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Region

Region	Count
Southeast	38
Heartland/Southwest	26
Northeast	49
Pacific	29
Midwest	19

The denominators for each of the below measures are the total respondents by region as depicted above.

Performance Measure 3a: (Provider Type) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Provider Type

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	25/38 (65.8%)	22/26 (84.6%)	31/49 (63.3%)	21/29 (72.4%)	15/19 (78.9%)
Nurse Practitioner	8/38 (21.1%)	4/26 (15.4%)	13/49 (26.5%)	7/29 (24.1%)	3/19 (15.8%)
Physician Assistant	2/38 (5.3%)	0/26 (0.0%)	5/49 (10.2%)	1/29 (3.5%)	1/19 (5.3%)
Other Providers	3/38 (7.9%)	0/26 (0.0%)	0/49 (0.0%)	0/29 (0.0%)	0/19 (0.0%)

Performance Measure 3b: (Provider Specialty/Subspecialty) Number of Providers in the SCDTDRCP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	18/38 (47.4%)	7/26 (26.9%)	17/49 (34.7%)	11/29 (37.9%)	11/19 (57.9%)
Hematology/Oncology	9/38 (23.7%)	16/26 (61.5%)	12/49 (24.5%)	12/29 (41.4%)	6/19 (31.6%)
Primary Care – Pediatrics	0/38 (0.0%)	0/26 (0.0%)	3/49 (6.1%)	1/29 (3.5%)	0/19 (0.0%)
Primary Care – Internal Medicine	1/38 (2.6%)	0/26 (0.0%)	1/49 (2.0%)	1/29 (3.5%)	1/19 (5.3%)
Primary Care – Family Medicine	2/38 (5.3%)	1/26 (3.9%)	2/49 (4.1%)	1/29 (3.5%)	0/19 (0.0%)
Primary Care – Med/Peds	1/38 (2.6%)	0/26 (0.0%)	1/49 (2.0%)	1/29 (3.5%)	0/19 (0.0%)
Hospitalist	2/38 (5.3%)	0/26 (0.0%)	0/49 (0.0%)	1/29 (3.5%)	0/19 (0.0%)
Emergency Medicine	0/38 (0.0%)	0/26 (0.0%)	1/49 (2.0%)	0/29 (0.0%)	0/19 (0.0%)
Other	5/38 (13.2%)	2/26 (7.7%)	10/49 (20.4%)	1/29 (3.5%)	1/19 (5.3%)

Performance Measure 3c: (Primary Practice Location-Rural/Urban Zip Codes) Number of Providers in the SCDTDRP Network Participating in Telementoring for Sickle Cell Disease (SCD) in the Past Year by Location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	0/38 (0.0%)	0/26 (0.0%)	0/49 (0.0%)	3/29 (10.3%)	1/19 (5.3%)
Urban	38/38 (100.0%)	26/26 (100.0%)	41/49 (83.7%)	26/29 (89.7%)	18/19 (94.7%)

Performance Measure 3d: (Regional/State Leads) Number of Providers in the SCDTDRCP Network Who Participated in Telementoring for Sickle Cell Disease (SCD) in the Past Year

Region	Numerator	Denominator	Percentage
Southeast	12	38	31.6%
Heartland/Southwest	7	26	26.9%
Northeast	12	49	24.5%
Pacific	11	29	37.9%
Midwest	6	19	31.6%

Performance Measures 4: (Total) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Region

Region	Numerator	Denominator	Percentage
Southeast	57	79	72.2%
Heartland/Southwest	46	73	63.0%
Northeast	48	61	78.7%
Pacific	34	43	79.1%
Midwest	36	50	72.0%

Performance Measure 4a: (Provider Type) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Provider Type

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	38/54 (70.4%)	40/66 (60.6%)	33/42 (78.6%)	25/32 (78.1%)	32/42 (76.2%)
Nurse Practitioner	16/19 (84.2%)	6/7 (85.7%)	10/14 (71.4%)	8/10 (80.0%)	3/7 (42.9%)
Physician Assistant	1/3 (33.3%)	0/0 (0.0%)	5/5 (100.0%)	1/1 (100.0%)	1/1 (100.0%)
Other Providers	2/3 (66.7%)	0/0 (0.0%)	0/0 (0.0%)	0/0 (0.0%)	0/0 (0.0%)

Note: The numerator is the number of providers by provider type in each region who report feeling comfortable treating sickle cell patients. The denominator is the total number of provider type by region

Performance Measure 4b: (Provider Specialty/Subspecialty) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	26/28 (92.9%)	13/14 (92.9%)	18/19 (94.7%)	12/15 (80.0%)	12/12 (100.0%)
Hematology/Oncology	19/24 (79.2%)	26/46 (46.5%)	13/17 (76.5%)	17/21 (81.0%)	17/21 (81.0%)
Primary Care – Pediatrics	1/3 (33.3%)	1/2 (50.0%)	3/3 (100.0%)	1/1 (100.0%)	0/6 (0.0%)
Primary Care – Internal Medicine	2/4 (50.0%)	0/1 (0.0%)	0/2 (0.0%)	1/1 (100.0%)	1/1 (100.0%)
Primary Care – Family Medicine	2/5 (40.0%)	1/2 (50.0%)	1/2 (50.0%)	1/1 (100.0%)	2/3 (66.7%)
Primary Care – Med/Peds	1/3 (33.3%)	0/1 (0.0%)	1/1 (100.0%)	1/1 (100.0%)	0/3 (0.0%)
Hospitalist	1/4 (25.0%)	0/0 (0.0%)	0/0 (0.0%)	1/1 (100.0%)	0/0 (0.0%)
Emergency Medicine	1/1 (100.0%)	3/3 (100.0%)	2/2 (100.0%)	0/0 (0.0%)	1/1 (100.0%)
Other	4/7 (57.1%)	2/4 (50.0%)	9/12 (75.0%)	0/2 (0.0%)	3/3 (100.0%)

Note: The numerator is the number of providers by specialty in each region who report feeling comfortable treating sickle cell patients. The denominator is the total number of provider specialty by region.

Performance Measure 4c: (Practice Location- Rural/Urban Providers- Zip codes) Number of SCDTDRCP Providers that Report Feeling Comfortable Treating Sickle Cell Patients by Location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	0/0 (0.0%)	1/2 (50.0%)	0/0 (0.0%)	2/3 (66.7%)	2/3 (66.7%)
Urban	57/79 (72.2%)	45/71 (63.4%)	32/40 (80.0%)	32/40 (80.0%)	34/47 (72.3%)

Note: The numerator is the number of providers by provider location in each region who report feeling comfortable treating sickle cell patients. The denominator is the total number of provider location by region.

Domain: Coordination and Delivery of Services (Hydroxyurea)

Performance Measures 5: (Total) Number of SCDTDRCP providers that saw at least one sickle cell patient in the past year, that prescribed hydroxyurea by Region

Region	Count
Southeast	55
Heartland/Southwest	43
Northeast	50
Pacific	18
Midwest	33

Performance Measure 5a: (Provider Type) Number of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient by provider type

Provider Type	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Medical Doctor	37/54 (68.5%)	38/66 (57.6%)	30/42 (71.4%)	15/32 (46.9%)	28/42 (66.7%)
Nurse Practitioner	16/19 (84.2%)	5/7 (71.4%)	13/14 (92.9%)	2/10 (20.0%)	4/7 (57.1%)
Physician Assistant	1/3 (33.3%)	0/0 (0.0%)	4/5 (80.0%)	1/1 (100.0%)	1/1 (100.0%)
Other Providers	1/3 (33.3%)	0/0 (0.0%)	0/0 (0.0%)	0/0 (0.0%)	0/0 (0.0%)

Note: The numerator is the number of provider type in the SCDTDRCP network (table above) that saw at least one sickle cell patient in the last year and prescribed HU to a sickle cell patient. The denominator is the total N of specified provider type.

Performance Measure 5b: (Provider Specialty/Subspecialty) Percent of providers in the SCDTDP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient by specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	26/28 (92.9%)	11/14 (78.6%)	18/19 (94.7%)	5/15 (33.3%)	11/12 (91.7%)
Hematology/Oncology	17/24 (70.8%)	27/46 (58.7%)	16/17 (94.1%)	9/21 (42.9%)	18/21 (85.7%)
Primary Care – Pediatrics	1/3 (33.3%)	1/2 (50.0%)	3/3 (100.0%)	1/1 (100.0%)	0/6 (0.0%)
Primary Care – Internal Medicine	2/4 (50.0%)	1/1 (100.0%)	0/2 (0.0%)	1/1 (100.0%)	1/1 (100.0%)
Primary Care – Family Medicine	2/5 (40.0%)	1/2 (50.0%)	2/2 (100.0%)	0/1 (0.0%)	0/3 (0.0%)
Primary Care – Med/Peds	2/3 (66.7%)	0/1 (0.0%)	0/1 (0.0%)	0/1 (0.0%)	0/3 (0.0%)
Hospitalist	1/4 (25.0%)	0/0 (0.0%)	0/0 (0.0%)	1/1 (100.0%)	0/0 (0.0%)
Emergency Medicine	0/1 (0.0%)	1/3 (33.3%)	0/2 (0.0%)	0/0 (0.0%)	0/1 (0.0%)
Other	4/7 (57.1%)	1/4 (25.0%)	4/12 (33.3%)	1/2 (50.0%)	3/3 (100.0%)

Note: The numerator is the number of specialty type in the SCDTDRCP network that saw at least one sickle cell patient in the last year and prescribed HU to a sickle cell patient. The denominator is the total N of specialty type.

Performance Measure 5c: (Provider Location - Rural/Urban Zip Code Providers) Percent of providers in the SCDTDRCP network that saw at least one sickle cell patient in the last year that prescribed hydroxyurea to a sickle cell patient by location

Provider Location	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Rural	0/0 (0.0%)	1/2 (50.0%)	0/0 (0.0%)	2/3 (66.7%)	0/3 (0.0%)
Urban	55/79 (69.6%)	42/71 (59.2%)	29/40 (72.5%)	16/40 (40.0%)	33/47 (70.2%)

Note: The numerator is the number of provider type in the SCDTDRCP network that saw at least one sickle cell patient in the last year and prescribed HU to a sickle cell patient. The denominator is the total N of provider location type (rural or urban).

Performance Measures 6: (Total) Number of sickle cell patients seen by a SCDTDRCP Network Provider wrote a hydroxyurea prescription in the past year by region

Region	Count	Total Patients	Percent
Southeast	11,798	12,265	96.2%
Heartland/Southwest	1,756	4,297	40.9%
Northeast	3,816	4,208	90.6%
Pacific	1,015	2,257	45.0%
Midwest	1,449	2,685	54.0%

Performance Measure 6a: (Age) Number of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year by population

Population	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Adult (≥18 years of age)	6,415/6,603 (97.2%)	692/2,225 (31.1%)	2,885/3,166 (91.1%)	437/1,042 (41.9%)	303/505 (60.0%)
Pediatric (<18 years of age)	5,383/5,662 (95.1%)	1,064/2,072 (51.4%)	931/1,042 (89.4%)	578/1,215 (47.6%)	1,146/2,180 (52.6%)

Note: The numerator is the number of SCD patients in population group seen by a SCDTDRCP provider in past year and received a prescription for HU in the past year. The denominator is the total N of population type.

Performance Measure 6b: (Provider Specialty/Subspecialty) Number of sickle cell patients seen by a SCDTDRCP Network Provider that had a hydroxyurea prescription in the past year by specialty

Provider Specialty/Subspecialty	Southeast N/D (%)	Heartland/Southwest N/D (%)	Northeast N/D (%)	Pacific N/D (%)	Midwest N/D (%)
Hematology	7,958/8,133 (97.9%)	550/1,968 (27.9%)	1,932/1,932 (100.0%)	303/910 (33.3%)	702/1,138 (61.7%)
Hematology/Oncology	2,463/2,575 (95.7%)	1,114/1,942 (57.4%)	1,174/1,174 (100.0%)	640/980 (65.3%)	561/1,280 (43.8%)
Primary Care – Pediatrics	3/11 (27.3%)	5/8 (62.5%)	66/66 (100.0%)	3/110 (2.7%)	0/2 (0.0%)
Primary Care – Internal Medicine	170/181 (93.9%)	10/10 (100.0%)	0/1 (0.0%)	60/200 (30.0%)	50/55 (90.9%)
Primary Care – Family Medicine	15/19 (79.0%)	45/94 (47.9%)	77/77 (100.0%)	0/40 (0.0%)	0/3 (0.0%)
Primary Care – Med/Peds	102/102 (100.0%)	0/8 (0.0%)	0/5 (0.0%)	0/6 (0.0%)	0/4 (0.0%)
Hospitalist	510/550 (92.7%)	0/0 (0.0%)	0/0 (0.0%)	8/10 (80.0%)	0/0 (0.0%)
Emergency Medicine	0/60 (0.0%)	2/141 (1.4%)	0/48 (0.0%)	0/0 (0.0%)	0/13 (0.0%)
Other	577/634 (91.0%)	30/126 (23.8%)	512/850 (60.2%)	1/1 (100.0%)	136/190 (71.6%)

Note: The numerator is the number of SCD patients seen by SCDTDRCP specialty provider in past year and received a prescription for HU in the past year. The denominator is the total N of specialty provider type.

Appendix A: Performance Measures (PM)/Provider Survey: Lessons Learned/Challenges

The NCC played a key role in aligning the provider survey with the performance measures data dictionary to ensure that the requested information was collected, aggregated and submitted to CoLab. The NCC worked closely with HRSA who led the development of approved the final performance measure domains and survey. Through an iterative process of review between the NCC, the RCCs, and HRSA, alignment was achieved and shared with the RCCs for dissemination. Distributed materials included a final performance measure provider survey, corresponding performance measure data dictionary, and provider network definition guidance. RCCs were responsible for survey distribution and aggregated regional data entry into CoLab.

There has been much to learn from this second implementation of the provider survey and collection of data. Below are some lessons learned and areas to refine related to the collection of performance measure data.

Lesson 1: Standard Definition of Who to Disseminate the Survey is Essential

Creating a standard definition ensured that each RCC more clearly understood and consistently applied the survey dissemination strategy. Finalizing this document was challenging, but importantly it assisted in streamlining the data collection approach.

Lesson 2: Implementation of Survey During a Pandemic

While clinical staff always have competing demands, during the time of COVID-19 this was amplified. RCCs reported that clinics were facing "organized chaos" with SCD clinics being transformed to meet critical pandemic related needs and staff having to face many unknowns but be responsive to multiple directives. Thus, capacity challenges were of great concern among most RCCs – both for the providers completing the survey and for RCCs themselves. This may have impacted survey distribution and response.

Lesson 3: Further Engagement with Rural Providers and Providers Outside of Academic Institution is Important to Having a Comprehensive National Picture of SCD Care

The sample of rural providers and providers outside of academic institutions was small. This may indicate the need to further engage with providers in these settings to be assured of a comprehensive picture of SCD care across the nation.

Lesson 4: In Addition to HU, Other Measures Should be Collected to Better Understand Access to Care for this Population

Responses from this survey reflect strong HU prescription rates. HU rates have historically been an important proxy for measuring access to care. However, SCD clinical care has evolved and this treatment is only one aspect of care. Going forward, including additional measures that capture other care coordination efforts, such as transitions to adult care, may enrich the understanding of access to care.

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Section 13 | State Plans: Pacific RCC, Heartland/Southwest RCC

Pacific RCC Action Plan

- Arizona State Action Plan
- California State Action Plan
- Colorado State Action Plan
- Nevada State Action Plan
- Oregon State Action Plan
- Washington State Action Plan

Heartland/Southwest RCC Action Plan

- Arkansas State Action Plan
- Iowa State Action Plan
- Kansas State Action Plan
- Louisiana State Action Plan
- Missouri State Action Plan
- Nebraska State Action Plan
- Oklahoma State Action Plan
- Texas State Action Plan

Section 14 | Qualitative Moderator Guides: Local Sites and RCCs

Digital Versions: Interview Guide: Local Site Leads | Interview Guide: RCC Leads

SCDTDRCP Stakeholder Interview Guide: Local Site leads	
Thank you agreeing to meet with us today. My name is [NAME] and I am a [ROLE] with the National Institute for Children's Health Quality also called NICHQ. I will be conducting the interview today. With me is my colleague [NAME], a [ROLE]. NICHQ is the National Coordinating Center (NCC) for the SCDTDP, funded by HRSA, which you participate within the [REGION'S NAME] region. Our work is focused on data collection from the regions and reporting to HRSA. The NCC is also responsible for producing a Congressional Report about the impact of the project. The purpose of this interview is to gather information about your site's programs and activities as part of the SCDTDP. Your participation in this interview will provide context and the content for the Congressional Report mandated by the 2018 legislation that funds SCDTDP. Today, we will discuss your site's work, including successes, challenges, best practices and lessons learned. This interview will last about 45 minutes. Your participation is completely voluntary; you can decline to answer any questions without any impact on your site as you work with [NAME OF REGIONAL LEAD] on SCDTDP. You can also choose to end the call at any time. Do you have any questions about NICHQ, the Congressional Report or this interview? *[respond to questions]* Great, before we begin, I want to let you know that we will be taking notes throughout the discussion. I would also like to record our session today to ensure we capture your responses correctly in our notes. NICHQ staff are the only ones who would listen to the recording. Do we have your permission to record the interview? ○ *[If yes]:* Thank you! ***[START RECORDING]*** ○ *[If no]:* Certainly. We will not record the interview.	
Questions	**Notes**
1) To begin, please tell me about your site, generally, things like: a. Where is your site located? i. Rural/urban b. Staffing question: i. ***[If speaking to a clinic]*** Configuration of your clinical staff who work with SCD patients? 1. Multidisciplinary vs. something else ii. ***[If speaking to a non-clinic]*** Describe the structure of your organization.	**Question Purpose:** Capture overview of local site organization, resources, capacity to do the work
c. What are the demographics of your patient population? i. Medicaid/medicare vs. private insurance d. What are your site's clinic/organizational priority areas regarding SCD work? e. ***[If a clinic]*** Does your site use an EMR (if so, which one) or conduct manual chart review for data collection? **Probe as needed to build out context for detail of interest:** • Tell us more about how and/or resources for collect and report data to your regional lead? • Tell us more about how your site is organized/staffed the SCDTDP project. • Tell us more about. . . Before leaving this question ask: • Anything else you'd like to add?	
2) Please describe how you work with your Regional lead. I am interested in items such as: • How do you access support if you need it? • The frequency of meetings with the regional lead • Decision-making process, etc. Before leaving this question ask: • Anything else you'd like to add?	**Question Purpose:** Engagement with regional collaborative structure and RCC. Relationship and benefits of having a backbone organization from local site perspective
3) I am interested in your site's SCDTDP QI projects. Please tell me about them. Probes: • How did you decide which QI projects to focus on? • Did you decide with the RCC, or did your site decide and let the RCC know what you were doing? • *[**If a site does not talk about HU work**, bring up as this is supposed to be addressed across all sites.]* To refer to as needed: **Project aims:** • Number of providers giving care • Number of patients receiving care • HU prescription (PM/QI) • Number of providers involved with Project ECHO (knowledge) **Clinical Objective(s) Quality Improvement area(s) of focus:** • HU Use (all sites and regions) • TCD screening (if eligible) Optional • Immunization (QI) Optional • transitions in care (QI) Optional	**Question Purpose:** Capture overview of RCC structure and organization.

• ECHO (QI) Optional Before leaving this question ask: • Anything else you'd like to add?	
4) Within your QI projects, what area(s) has (have) been your site's biggest successes? What helped make it a success? For purposes of this question success is how you, the local site lead and staff, define it. **Probe as needed to build out detail of interest:** a. Can you tell me more about . . . ? b. How did structure/resources facilitate success? c. Are their next steps coming out of your successful initiatives? d. How would you describe best practices related to your success? Before leaving this question ask: • Anything else you'd like to add?	**Question Purpose:** Capture RCC attributed successes and facilitators
5) Within your SCDTDP QI projects, what have been your site's biggest challenge(s)? **Probe as needed to build out detail of interest:** a. Can you tell me more about . . . ? b. What challenge(s) your site/state still faces? c. What might help to remove the barriers posed by challenges you continue to experience? Before leaving this question ask: • Anything else you'd like to add?	**Question Purpose:** Capture RCC attributed challenges, barriers and resolution
6) For your challenges, did you have any resolutions? Before leaving this question ask: • Anything else you'd like to add?	
7) ***[If site did not discuss Covid-19 impact]*** How has Covid-19 impacted your work for the SCDTDP project? a. Probe: have you increased your use of telemedicine with SCD patients? Before leaving this question ask: • Anything else you'd like to add?	
8) Besides what we have discussed is there anything else that you would like to highlight about your site's work for this project? Before leaving question ask: • Anything else you'd like to add?	**Question Purpose:** Capture additional input of importance from the RCCs perspective in relation to CR context

SCDTDRCP Stakeholder Interview Guide: RCC leads

Thank you for taking the additional time out of your schedule to meet with me today. Your participation in this interview will provide context and the content for the development of a Congressional Report, Model Protocol and Compendium of Resources all mandated by the 2018 legislation that funds the SCDTDRCP initiative.

This interview will last about 45 minutes. Your participation is completely voluntary; you can decline to answer any questions without any impact on your RCC's participation with SCDTDRCP. You can also choose to end the call at any time.

I will be taking notes throughout the discussion. I would also like to record our session today to ensure we capture your responses correctly in our notes. NICHQ staff are the only ones who would listen to the recording. Do we have your permission to record the interview?

- *[If yes]:* Thank you! ***[START RECORDING]***
- *[If no]:* Certainly. We will not record the interview.

As I previously introduced, I am going to ask questions about your experience as the RCC, including your **region's successes, best practices, and lessons learned as you have been addressing some or all of the programs objectives which include:**

By 2021, increase by 10 percent from baseline:

1. the total number of providers, including primary care providers, participating in **telementoring and telemedicine activities.**
2. **the number of providers treating individuals with sickle cell disease** in each state using the NHLBI Expert Panel Report recommended treatments and prevention.
3. the number of eligible individuals with sickle cell disease receiving a **hydroxyurea prescription** at least twice in the past year among patients seen by participating providers.
4. the number of individuals with sickle cell disease seen at participating institutions that have documented recommended **pneumococcal vaccinations at least annually.**
5. the number of eligible individuals with sickle cell disease seen at participating institutions that have documented **Transcranial Doppler Ultrasound (TCDs) at least annually**.
6. the number of eligible adolescents with sickle cell disease seen at participating institutions that have a **documented transition plan**.

Before I begin with the interview, do you have any questions?

Questions	Notes
1) To begin, please describe the structure of your RCC. I am looking for a general overview of how you communicate/manage the work that is done within your region. **Probe as needed to build out context for detail of interest:** • Do you have monthly calls as a group? • Do you have individual calls with each site? • Do your local sites reach out to you, or do you initiate the contact? • How are decision made re: choosing QI projects? • How do you communicate changes or things that all sites need to know?	**Question Purpose:** Capture overview of RCC structure and organization.
2) Please describe the QI initiatives in your region and who is responsible for overseeing the projects? **Probe:** • Which sites are conducting with initiatives?	**Question Purpose:** Capture breadth and/or variation of QI initiatives across region.
3) Among the project aims, what activity(ies) has (have) been your region's biggest success(es) in the project to-date? What helped make it a success? For purposes of this question success is how you, the RCC leads and staff, define it. • ***[As needed, share on screen during interview. Project aims were also emailed ahead of the interview.]*** **Probe as needed to build out detail of interest:** • Can you tell me more about . . . ?	**Question Purpose:** Capture RCC attributed successes and facilitators **Project aims:** • Number of providers giving care • Number of patients receiving care • HU prescription (PM/QI) • Number of providers involved with Project ECHO (knowledge) **Clinical Objective(s) Quality Improvement area(s) of focus:** • HU Use (all sites and regions) • TCD screening (if eligible) Optional • Immunization (QI) Optional • transitions in care (QI) Optional • ECHO (QI) Optional

4) Among the project aims, what has (have) been your region's biggest challenge(s) in the project to-date? What were the barriers and resolutions? **Probe as needed to build out detail of interest:** • Can you tell me more about . . . ?	**Question Purpose:** Capture RCC attributed challenges, barriers and resolution Related to aims and areas of focus in cell above
5) Are there one or two site leads that you recommend we speak with to gain additional information about bright spots in your Region's SCDTDRCP work?	**Question Purpose:** Capture RCC identification of site leads for NCC to interview
6) Besides the topics we have discussed is there anything else that you think is important to highlight in the Congressional Report about your region?	**Question Purpose:** Capture additional input of importance from the RCCs perspective in relation to CR context

Section 15 | Other Collaboratives with National SCD Organizations and Programs

Although not fully or specifically supported by resources from the Program, the network participated in multiple synergistic activities designed to improve care for people living with SCD. Described here are several far-reaching activities that RCCs participated in during this funding period, which were accomplished in addition to the primary work of this Program. The established nature of the network and the RCC commitment to participate were crucial in the success of these endeavors.

STAMP

The structure of Project ECHO® for provider education has worked well for SCD. And given the successes, the need to expand this educational opportunity is clear but remains challenging. Engaging PCPs is one strategy to increase the knowledgeable provider pool. However, in prior work, RCCs had seen limited involvement with PCPs in their ECHOs. In Fall 2018, with additional, limited funding from HRSA's Office of Minority Health (OMH), RCCs were asked to participate in a supplemental national Project ECHO® initiative: *Sickle Cell Disease Training and Mentoring Program (STAMP)*. This program was established at the behest of the Assistant Secretary for Health at HHS. All five of the Program RCCs participated in piloting this targeted ECHO to try and engage, educate, mentor, and facilitate clinical co-management support for PCPs.

Using the Project ECHO® model, the Northeast RCC team coordinated the efforts of this twice-monthly meeting. Recruitment and marketing were overseen by the Office of the Assistant Secretary of Health and the Office of Minority Health (OMH) and CME credit was provided by STORM to encourage participation. See Table 1 for a list of STAMP sessions. During a six-month period (January-June 2020), 12 sessions were held, which were attended by 546 providers (214 unique participants).

In summary, attendees from all sessions represented 33 U.S. states and 10 additional countries (Greenland, Ghana, India, Jamaica, Nigeria, Uganda, Tanzania, the United Kingdom, Canada, North Macedonia). Primary Care Providers (PCPs) and Advanced Practice Providers (APPs) — nurse practitioners and physician assistants — represented 22 U.S. states and Washington D.C., Canada, and Ghana. Experience and number of patients with SCD they were treating varied significantly but averaged 10 years of experience and 32 patients seen in the last 12 months. All were outpatient providers and some reported working in a Federally Qualified Health Center (FQHC) or look-a-like. There were 75 licensed prescribers (physicians, APPs). Overall, sessions were well attended, averaging 45.5 per session. No providers indicated practicing in a rural zip code. An assessment showed positive reactions to this Project ECHO® series: 100% noted they would come to a future Project ECHO® session and recommend STAMP ECHO to their colleagues. However, the denominator (n=8) was small and care should be taken with interpretation.

Despite the strong attendance, the main outcome of engaging PCPs did not see the level of demand desired in the target audience, and the telementoring was very limited — only one case was presented by STAMP registrants when given 24 opportunities. While the attendees did increase their knowledge and

Table 1. List of STAMP Sessions During a Six-Month Period

1/8/2020	12:00 PM, EST	Update on Pathophysiology of SCD	Sophie Lanzkron, MD, MHS	SiNERGe
1/16/2020	2:00 PM, EST	Hydroxyurea for Adults	Russell Ware	STORM
2/4/2020	1:00 PM, CST	Imaging Uncomplicated Headache in SCD	James Harper	Heartland | Southwest
2/18/2020	4:00 PM, CST	Screening Assessments in SCD	Julie Kanter, MD	EMBRACE
3/5/2020	12:00 PM, PST	Transfusion in SCD	Trisha Wong	Pacific
3/20/2020	12:00 PM, EDT	New Medications for Sickle Cell Anemia	Charles Quinn, MD MS	STORM
4/7/2020	5:00 PM, EDT	Screening Assessments in SCD	JJ Strouse, MD, PhD	EMBRACE
4/29/2020	12:00 PM, EDT	Tips and Techniques for Pain Management in Sickle Cell Disease	Wally Smith, MD	SiNERGe
5/1/2020	12:30 PM, CDT	Self Management Techniques for Adults with SCD	Donna McCurry, APRN, FNP-BC	Heartland | Southwest
5/19/2020	11:00 AM, MDT	Back to Basics: Common Lab Findings in SCD	Kathryn Hassell, MD	Pacific
6/10/2020	12:00 PM, EDT	Contraceptives in SCD	Lydia Pecker, MD	SiNERGe
6/23/2020	4:00 PM, MDT	Telemedicine in SCD	Julie Kanter, MD	EMBRACE

holding these sessions was considered worthwhile, RCCs did not experience this as an effective way to reach and train PCPs. Additionally, the results of this focused project further reinforced the RCCs' strong belief that care should be centralized with an expert in SCD care. While PCPs should be integrated, placing heavy clinical responsibility on this group is not advised. In general, if PCPs provide care to people living with SCD in their patient panel, it is limited. Understandably, they have neither the time to keep up with evolving recommended care nor the bandwidth needed for this often-complex group of patients.

Turning Attention to Advanced Practice Providers

At the end of the STAMP pilot period, focus shifted from STAMP to initiating a Sickle Cell Disease Advanced Practice Providers Opportunities Resources and Training (sAPPort) with the RCCs in December 2020. As with STAMP, the OMH provided additional funding to support sAPPort, which is designed to specifically address educational needs of Advance Practice Providers (APPs) — nurse practitioners and physician assistants — a group that RCCs believe are key to developing a sustainable SCD clinician pipeline. The supplemental funding was allocated for a one-year implementation. As with STAMP, sAPPort is a national RCC effort. The Project ECHO® sessions began in Winter 2020 and included a didactic presentation on SCD care aspects, including Acute Chest Syndrome, neurological complications, ED management, and pain management. During each session, one case presented by an APP was discussed among participants and an expert APP panel. The Northeast and Southeast RCCs collaborated to support this monthly ECHO. All faculty are APPs to ensure that content is targeted and tailored. RCCs are also participating in other activities to ensure focused and comprehensive SCD education is provided to APPs across the country. RCCs are in the process of developing an asynchronous, online learning curriculum for APPs who wish to receive formal certification to treat people living with SCD. The Northeast region spearheaded this program with APP faculty who determined what content should be included in the curriculum. RCCs have identified SCD experts from across their regions to provide recorded lectures for this curriculum. Registered APPs will be required to go through all lectures and complete pre- and post-assessments and learning activities to pass the curriculum and receive certification. Additionally, each RCC identified one to two APPs within their region to be funded mentees for all aspects of the sAPPort program. These APPs regularly meet with SCD experts from their regions and are required to attend sAPPort ECHOs and complete the online curriculum. They will also be invited to complete observerships at SCD centers of excellence to get hands-on experience and mentorship in working with people living with SCD.

Collaboration with the CDC Sickle Cell Disease Data Collection Program

Begun in 2015, the Sickle Cell Data Collection (SCDC) program collects health information about people with SCD to study long-term trends in diagnosis, treatment, and healthcare access for people living with SCD in the United States. The program, currently being implemented in 11 states, helps to inform policy and healthcare standards that improve and extend the lives of people with SCD.

This program is helping participants to better understand the following areas:

- Where people with SCD live
- Transition from pediatric to adult SCD care
- Hispanic patients with SCD
- Older patients with SCD
- The use of healthcare services for SCD

Several of the Program states participate in this program, which will help the network build capacity through engaging local partnerships and across states.

RCC COVID-19 Telemedicine Survey

In 2020, the RCCs, using their established partnerships, met to discuss how to capture information about what was happening in the moment regarding telemedicine and COVID-19. Separate from the Program RCC responsibilities, all the RCCs worked together to plan and conduct a telemedicine survey and decide which quality improvement measures to track.

Long-term goals of telemedicine-related activities include:

- Assess feasibility of telemedicine from the provider and patient perspective

- Assess effectiveness in providing SCD care
- Evaluate barriers to care with telemedicine
- Evaluate use of telemedicine for SCD visits throughout the nation

Telemedicine activities consisted of three components:

1. Provider Survey

Survey 1: Assessing Organizational Readiness was the short-term goal of the initial survey, which was sent (5/16/20-6/16/20) to primary SCD providers, including MDs, Doctors of Osteopathic Medicine, APPs, social workers, and psychologists

Survey 2: Distributed 6 months post-baseline; when analyzed will assess feasibility, barriers, and positives/negatives of telemedicine use

Survey 3: Distributed 12 months post-baseline; when analyzed will assess feasibility, barriers, and positives/negatives of telemedicine use

2. Patient Survey

All patients who received a telemedicine visit within a month would be asked to complete:

- Survey 1: Baseline
- Survey 2: (6 months)
- Survey 3: (12 months)

Goal: Survey as many patients as possible, up to 50/period

Goal: Assess the feasibility of telemedicine for SCD from the patient perspective, including barriers/facilitators and positives/negatives of use

3. Quality Assessment Measures

RCC leads worked together to identify data metrics to track the use of telemedicine for patient visits across the nation

Each RCC site was asked to collect data on the following each month:

- Total clinic volume (number of appointments scheduled)
- Number of appointments scheduled and completed in person
- Number of appointments scheduled and completed via telemedicine
- Number of lab visits scheduled and completed for associated telemedicine visits

Goal: Determine the use and effectiveness of telemedicine for SCD patient visits

Data for applicable time points have been collected and analysis is forthcoming.

Work with the Sickle Cell Disease Association of America

The Sickle Cell Disease Association of America (SCDAA) is a well-known, trusted organization that focuses on addressing the needs of people living with SCD and their families in the U.S. and beyond. As COVID-19 was evolving, SCDAA, led by Dr. Biree Andemariam, SCDAA Chief Medical Officer, gathered a team of experts and formed the Medical and Research Advisory Committee (MERAC). Starting in 2020 during the COVID-19 pandemic, MERAC has posted key advisory statements and developed informational tools, letter templates, and infographic tips for both patients and caregivers. These information sources are publicly available online. Program clinics assembled specific education for patients using these resources.

Establishment of and Participation in the National Alliance of Sickle Cell Centers (NASCC)

In 2020, several of the Program PIs helped launch and are serving as presiding officers in this newly formed organization. The mission of this organization is to support SCD centers in delivering high-quality comprehensive care by setting standards of care and promoting their adoption, identifying opportunities and resources to strengthen SCD centers, and advocating for access to comprehensive care to improve health outcomes, quality of life, and survival.

Goals

- Create an infrastructure for adult and pediatric SCD centers to define, continually enhance, and promote the adoption of standards of primary and specialized care that comprise a comprehensive care center for people living with SCD
- Develop tools and share information with SCD centers to use in implementing and operating a comprehensive care model within their organizations

- Enable access to the Globin Research Network for Data and Discovery (GRNDaD), a multi-site registry used to optimize quality improvement and quality assurance through data analytics
- Identify opportunities and resources (federal, state, and private) that SCD centers can leverage to sustain funding and ensure equitable access to comprehensive care
- Create and sustain an advisory board of stakeholders committed to improving outcomes for people living with SCD
- Work with the American Society of Hematology (ASH) and other similar organizations to accomplish these goals and to collaborate on research, data collection, advocacy, and other efforts of mutual interest

In collaboration with the GRNDaD network, this group hosted a speaker series focused on SCD research:

Example Topics

- *Pathophysiology of white matter damage in Sickle Cell Disease: Challenges and Controversies*, John Wood, MD, PhD
- *Thick and Thin: Clotting and Bleeding in Patients with Sickle Cell Disease*, Ted Wun, MD, FACP
- *Red Cell Rheology: Biomarkers to evaluate sickle cell disease therapies,* Vivien Sheehan, MD, PhD

Participation Metrics

- Anyone was able to register, but attendance was mostly comprised of researchers, physicians, and APPs
- 18 sessions to date
- Total Attendance: 1,846
- Average attendance: 103 attendees per session (ranging from 70-156)

Section 16 | Model SCD Clinic Presentation

Digital Version: Model SCD Clinic Presentation

The EMBRACE (Education and Mentoring to BRing Access to CarE) Sickle Cell Disease Network 2021-25

John J. Strouse, MD, PhD
Associate Professor of Medicine and Pediatrics
Divisions of Adult Hematology and Pediatric Hematology/Oncology

THE EMBRACE NETWORK

Duke University School of Medicine

Partnerships with CBOs

- Models of partnership
  - State PI with individual CBOs
  - Lead CBO for region
  - National CBO partners
- HRSA Newborn Screening Follow-up Program
  - CBO partners involved in some states/regions

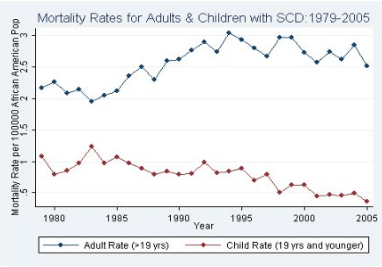
sickle cell 3 community consortium

The William F. Proudford Sickle Cell Fund, Inc.®

- Better health outcomes for affected individuals
- Care coordination throughout the medical system
  - Vertical
  - Horizontal
  - Continual bi- directional relationship with their medical team.
- Ensure appropriate access to disease modifying therapies
- Preventative care
- WHY: to improve quality of life for patients to achieve longer, better lifespans
- AND….Improved relations between patients and health care personnel
- AND….Improved financial outcomes for the institution

WIN WIN WIN

Mortality Rates: Adults and Children



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Outline

- What is a sickle cell center?
- What do you need to have (essentials) to be a SCD center
- What types of models of centers are there
- Examples of current centers
- NASCC

- A thoughtful, team-based care approach to adults living with sickle cell disease
- Consistent, comprehensive and coordinated care for adults with sickle cell disease throughout the "medical environment"
- A center with a programmatic emphasis on SCD for affected adults
- A center that coordinates SCD care

EMBRACE SCD Network

Multiple PIs: Osunkwo, Strouse, and Kanter-Washko

State	PI	Institution
Alabama	Brandi Pernell	UAB
Florida	Ofelia Alvarez	University of Miami
Georgia	Peter Lane/Betty Pace	Emory/ Augusta University
Kentucky	Ashok Raj	University of Louisville
Mississippi	Tobe Momah	University of Mississippi
North Carolina	Ify Osunkwo/JJ Strouse	Carolinas HealthCare System
South Carolina	Alan Anderson	Palmetto Health
Tennessee		University of Tennessee

- Sickle Cell Disease: a rare disease that still limits life expectancy through chronic complications despite childhood survival and availability of therapies
- At the individual level, it is about where you live:
  - Healthcare opportunities and insurance are state-based and even more local than that
  - Insurance may be available but ≠ access to necessary care
  - Best care often equilibrates to best advocates or knowledge

GOAL: Shift programmatic emphasis to how to build a system of care to enhance access and coordination of care (for affected adults)

The Essentials

- A programmatic, multidisciplinary, team-based approach to care
- The SCD center should also be the recognized authority (leader within their larger hospital or academic center) for managing SCD as a patient population

- Lead nurse/clinic manager who could also serve as the center director or co-director with the SCD specialist.
- This person helps share responsibility with the director for upkeep and implementation of guidelines and protocols as well as training and audits related to the center.
- The clinic manager can have additional responsibilities including quality improvement and oversight.
- Dedicated infusion space or a day hospital **
- Behavioral and mental health provider (psychologist) **
- Transition plan for helping children adapt and transfer to the adult care setting
- Access to a gynecology provider or other means of providing contraception for women with SCD

** longer to reach consensus

- The centers surveyed were also categorized into different models defined based on the size of the center, patient population, structure (within a division or department), services provided (whether they had primary care within the SCD center), and staffing for care delivery (
- These models were also defined by their location of services and whether they are stand-alone clinics or embedded within other divisions/departments/clinics

- SCD Comprehensive Care Clinic (Stand alone)
- Embedded Adult SCD Program
- Specialized SCD medical home

+Hub and Spokes Model of care

* What is an affiliate (spoke)

- A physician lead who is considered a SCD specialist
  - This individual will accept responsibility for establishing protocols for care, training, implementing audits, and share in the overall responsibility for the management of the clinic.
  - The lead SCD specialist should be comfortable providing evidence-based pain management, should undertake continuing professional development of relevance to this role, and have an established plan for how to cover for absences.
- One or more social workers
- A patient coordinator (sometimes called a patient navigator or case manager)
- Dedicated nursing staff
- Ability to offer acute and chronic pain management
- Transfusion (including apheresis)
- Timely access to specialist services.

Adjunct

- What type of center will you be:
  - Ambulatory care center vs. dedicated infusion center/clinical care
- How big is your population?
- How big is your team (and who is missing from your center model and what is the timeline?)
- What resources are currently available?
- Hours of operation, adapting for various circumstances (critical pts, workflow and chain of command)
- Plan to allow for expansion

MUST HAVE:

A plan to provide Team-Based Care for adults with SCD

Institutional support

SHOULD HAVE:

- A business plan (at least in the beginning)
- A plan for community-based organization interaction
- A plan for quality assessment and improvement

- Preferred but not necessary
- Primary care provider
- A physical therapist
- Occupational therapist
- A pharmacist
- Accessible dental care (as may be available hemophilia treatment centers
- SCD educator.

- **Provides excellent team-based SCD Care**
- Dedicated clinical space and staff
- Includes an infusion center/day hospital
- Led by a sickle cell specialist
- Has access to all necessary subspecialists to provide true comprehensive care
- Urban model where the focus is not on reaching an outside population
- Has a plan for primary care and co-management with PCP
- Includes mental health within the SCD care model

Optimal

Models of sickle cell centers

- Partnership model-often within the Cancer Center (but other options)
- Shares clinical space and personnel with Oncology
- **Provides excellent team-based SCD Care**
- Often shares an infusion center/day hospital
- Led by a sickle cell specialist but has multiple team members with hematologic experience and practice
- Has access to all necessary subspecialists but may not have them directly part of the SCD center

- Dedicated clinical space and staff for SCD
- **Provides excellent team-based SCD Care**
- **Includes primary care within the medical home**
- **Must have a care coordinator within the medical home**
- May/may not include an infusion center/day hospital (but must have a plan to provide necessary pain management)
- Led by a sickle cell specialist
- Suburban or urban model where individuals may come in from further away or have difficulty coordinating care

- SCD centers for adults must provide compassionate, comprehensive care that is coordinated to ensure continuous and personalized care with attention to both the physical and emotional well-being of the individual.
- Sickle cell disease must be lead by the sickle cell center within the greater institution
- Providing standardized, **equitable** patient-centered management will ensure higher quality, more cost-effective care for this vulnerable patient population.

Thank you!!!!

- **American Society of Hematology**
- My workshop co-leads: Wally Smith, Sophie Lanzkron, John Roberts
- Co-authors: Payal Desai, Marsha Treadwell, Biree Andemariam, Jane Little, Diane Nugent, Susan Claster, Deepa Manwani, John Strouse, Ifeyinwa (Ify) Osunkwo, Rosalyn Stewart, Allison King, Lisa Shook, Judith Baker
- Blood Advances for publishing "Building access to care in adult sickle cell disease: defining models of care, essential components, and economic aspects " today
- NASCC CO-FOUNDERS:
  - Sophie Lanzkron, Marsha Treadwell, Kim Smith-Whitley, Deepa Manwani

- **The Hub can be a part of any of the above centers**
  - Led by a sickle cell specialist
  - Has access to all necessary subspecialists
- The affiliate spokes:
  - MUST be affiliated with the Hub
  - MUST have a plan for supervision and emergency management
- Who can run an affiliate?
  - Primary care physicians with contacts to the hub
  - Specialized Advanced Care Providers
  - May have SCD infusion areas or (more likely) shared infusion space in which they can provide pain management
  - Must include telehealth/outreach with the HUB

National alliance of sickle cell centers

Presented by:

- Options for all types of modifying therapy: hydroxyurea, glutamine, voxeletor, crizanlizumab
- SCD prevention assessments
- Iron overload management program
- Blood transfusion management avoid inappropriate blood transfusion, extended cross-matching of blood,
- Individualized pain management plan/program
- Family planning plan
- Mental health plan

- **Mission:** To support sickle cell disease (SCD) centers in delivering high-quality comprehensive care by setting standards of care and promoting their adoption, identifying opportunities and resources to strengthen SCD centers, and advocating for access to comprehensive care to improve health outcomes, quality of life, and survival.
- **Goals**
- Create an infrastructure for adult and pediatric SCD centers to define, continually enhance, and promote the adoption of standards of primary and specialized care that comprise a comprehensive care center for adults and children with SCD.
- Develop tools and share information with SCD centers to use in implementing and operating a comprehensive care model within their organization.
- Enable access to the Globin Research Network for Data and Discovery (GRNDaD), a multi-site registry to optimize quality improvement and quality assurance through data analytics
- Identify opportunities and resources (federal, state, and private) that SCD centers can utilize to sustain funding and ensure equitable access to comprehensive care.
- Create and sustain an advisory board of stakeholders committed to improving outcomes in SCD
- Work with the American Society of Hematology, and other similar organizations to accomplish these goals and to collaborate on research, data collection, advocacy, and other efforts of mutual interest.

- Telehealth: used to provide direct PROVIDER to Patient Care
  - Enhance hub-spoke model of care
  - Can be used to help provide inpatient care
- Telementoring (ECHO)
  - increasing local physician knowledge and self-efficacy in managing individuals with SCD (based on the ECHO program)
- Extended Hours
  - Hours outside of typical 8-5 clinic hours used to provide additional pain management or off-hours clinic management
- ER Observation Units
  - Partnership with ER facility to provide extended care (that does not require admission)

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Appendix C: Data Methodology

REPORT TO CONGRESS
SEPTEMBER 2021

Data and Measurement Methods

Data collection of components of care for people living with Sickle Cell Disease (SCD) was a foundational component of the Program. Periodic data collection of care measures ensured that grantees focused on the Program priorities that were set forth in the 2017 HRSA/MCHB FOA.

This section offers detail regarding the following areas:

- The groups involved with the data collection
- The methodology for each of the data collection streams
- Reflections about future data collection

Regional Coordinating Centers and National Coordinating Center Partnership

The HRSA/MCHB funded five RCCs under a cooperative agreement. HRSA contracted with the National Institute for Children's Health Quality to serve as the NCC to coordinate data collection and analyze data. The RCCs and the NCC were funded separately to conduct complementary work for the Program.

The RCCs provided regional and local data and materials to the NCC for the data collection activities.

The NCC's major areas of work were collection, aggregation, and analysis of data from the RCCs and development and collection of materials for this Congressional Report, providing Congress and the public the results of their federal investment focused on improving the health and lives of people living with SCD.

The NCC was also contracted to convene an Oversight Steering Committee (OSC). The OSC was comprised of RCC leads and additional experts who brought specific knowledge, skills, and connections to assist in making recommendations to the Program. OSC members provided input on measurements and gave additional expert feedback and updates about SCD work. See Table 1 for the roles of these three groups and Table 2 for information on RCC lead institution(s), their participating sites' information, and their participating community-based organizations' information.

Roles of Data Development and Collection Roles of the NCC, RCCs, and OSC

This section details the specific tasks related to the NCC's data management and collection role as well as the RCCs' data responsibilities.

The NCC, in collaboration with HRSA/MCHB and the RCCs, created a collective data and measurement plan for the implementation and collection of the Program measures. The NCC supported the fielding and implementation of two data streams, described below, by finalizing uniform data definitions for the data dictionary. The NCC supported and provided technical assistance to RCCs as they performed ongoing data collection and submission. The NCC was also responsible for aggregating regional data to provide nationally comprehensive data reports and disseminating findings across regions and to HRSA.

Table 1. Data Collection-Focused Activities During the 2017-2021 Program

DATA COLLECTION-FOCUSED ACTIVITIES		
NATIONAL COORDINATING CENTER (NCC)	REGIONAL COORDINATING CENTERS (RCCS)	OVERSIGHT STEERING COMMITTEE (OSC)
• Finalized data dictionary and manual of operating procedures (MOP) for both Provider Survey for Performance Measurement (PSPM) and Clinical Quality Improvement Measures (CQIM) • Organized and facilitated monthly Data Management Working Group (DMWG) meetings attended by all 5 RCCs and HRSA/MCHB • Organized and facilitated monthly meetings with each of the five RCCs to share challenges and solutions in data collection efforts • Organized and facilitated OSC meetings twice a year • Provided ongoing technical assistance to RCCs • Collected, aggregated, and developed an annual report for PSPM data • Collected, aggregated, and developed a quarterly report for CQIM data • Provide clinical and medical informatics expertise	• Contributed clinical expertise to measure set development • Fielded annual PSPM and compiled data for NCC analysis • Collected and submitted CQIM data quarterly to NCC for analysis and reporting • Alerted the NCC of concerns and anomalies and annotated issues in the data set submitted in NICHQ's CoLab	• Provided balanced guidance and objective advice to shape Program activities and guide implementation • Ensured activities were aligned with the objectives listed in the legislation as well as the specific aims of the Program • Provided timely knowledge about current trends as well as identified upcoming political, legislative, and regulatory developments that could affect the work • Provided opinions and assistance as requested in evaluating relevant data and preparation of required annual reports

Figure 1. Map of National Coordinating Center, Regional Coordinating Centers, Clinics and Participating Sites, and Community-Based Organizations

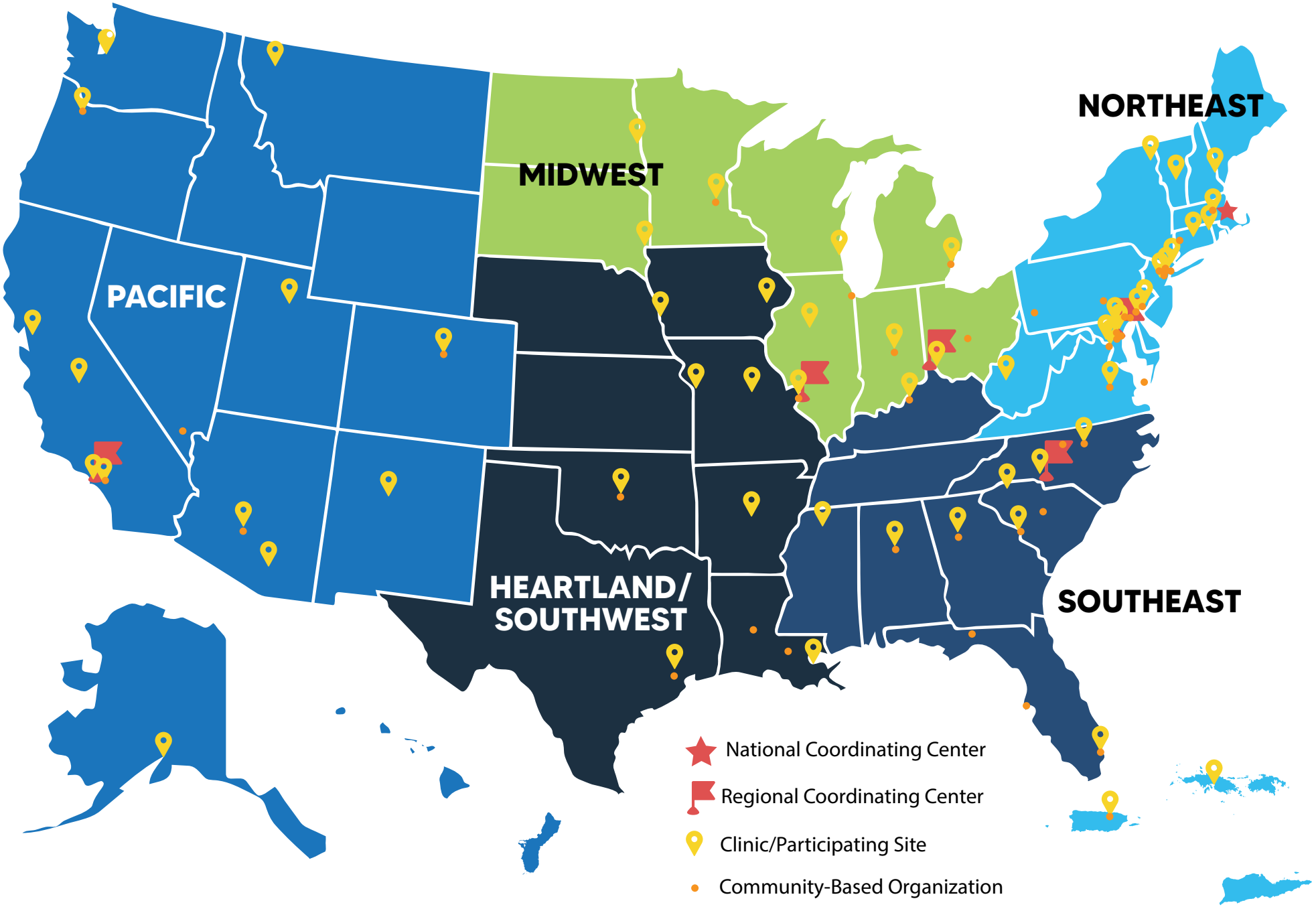


Table 2. Participating Sites and Community-Based Organizations within Regional Coordinating Centers

RCC	LEAD ORGANIZATION	STATES TERRITORIES	PARTICIPATING SITES	COMMUNITY BASED ORGANIZATIONS
	**Center for Inherited Blood Disorders (CIBD)**	**AK, AZ, CA, HI, ID, NV, OR, Pacific Basin, WA**	**1.** AK – Alaska Pediatric Oncology **2.** AZ – University of Arizona Health Sciences Center; Phoenix Children's Hospital **3.** CA – Center for Inherited Blood Disorders; UCSF Benioff Children's Hospital Oakland; Martin Luther King, Jr. Outpatient Center, Valley Children's Hospital **4.** CO – Colorado Sickle Cell Disease Treatment And Research Center, University of Colorado **5.** NM – University of New Mexico **6.** MT – Kalispell Regional Healthcare **7.** NV – Sickle Cell Center of Nevada **8.** OR – Oregon Health and Science University **9.** WA – Odessa Brown Children's Clinic, Seattle Children's Hospital **10.** UT – Utah Center for Bleeding and Clotting Disorders at Primary Children's Hospital	**1.** Sickle Cell Disease Foundation of California **2.** Bridging The Gap - Adult Sickle Cell Disease Foundation of Nevada **3.** Dreamsickle Kids Foundation (Nevada) **4.** Sickled Not Broken Foundation of NV **5.** The Sickle Cell Foundation of Arizona **6.** Colorado Sickle Cell Association **7.** Sickle Cell Anemia Foundation of Oregon, Inc.
	Cincinnati Children's Hospital Medical Center	IL, IN, MI, MN, ND, OH, SD, WI	**1.** IL – Children's Hospital of Illinois-Peoria **2.** IN – Indiana Hemophilia & Thrombosis Center **3.** MI – Sickle Cell Disease Association of Michigan **4.** MN – Children's Minnesota **5.** ND – Sanford Health (Fargo) **6.** OH – Cincinnati Children's Hospital Med Center **7.** SD – Sanford Health (Sioux Falls) **8.** WI – Children's Hospital of Wisconsin	**1.** Sickle Cell Disease Association of America – Michigan Chapter **2.** Sickle Cell Disease Association of America – Illinois Chapter **3.** The Martin Center (Indianapolis, IN) **4.** Sickle Cell Foundation of Minnesota (Minneapolis, MN) **5.** Ohio Sickle Cell and Health Association (Columbus, OH)
	Washington University School of Medicine, St. Louis	AR, IA, KS, LA, MO, NE, OK, TX	**1.** AR – University of Arkansas for Medical Sciences, Arkansas Children's Hospital and Research Institute **2.** IA – University of Iowa Stead Family Children's Hospital **3.** KS – University of Kansas Medical Center **4.** LA – Louisiana State University Pediatrics, New Orleans Children's Hospital **5.** MO – Washington University School of Medicine, Barnes Jewish Hospital, St. Louis. Children's Hospital, Truman Medical Center, Missouri University Health Care **6.** NE – University of Nebraska Medical Center, Children's Hospital and Medical Center **7.** OK – University of Oklahoma Health Sciences Center **8.** TX – Baylor College of Medicine, Texas Children's Hospital	**1.** The Sickle Cell Association (St. Louis) **2.** St. Louis Integrated Health Network **3.** Sickle Cell Association of South Louisiana and Baton Rouge Sickle Cell Anemia Foundation **4.** Supporters of Families with Sickle Cell Disease (Oklahoma) **5.** Sickle Cell Association of Texas, Marc Thomas Foundation

RCC	LEAD ORGANIZATION	STATES TERRITORIES	PARTICIPATING SITES	COMMUNITY BASED ORGANIZATIONS
SOUTHEAST 	Levine Cancer Institute, Atrium Health	AL, FL, GA, KY, MS, NC, SC, TN	**1. AL – University of Alabama at Birmingham** **2. FL – University of Miami** **3. GA – Augusta University, Emory University/CHOA** **4. KY – University of Louisville** **5. MS – University of Mississippi** **6. NC – Duke University, Atrium Health** **7. SC – Prisma Health**	**1.** Piedmont Health Services and Sickle Cell Agency (North Carolina) **2.** Bridges Pointe, Inc. Sickle Cell Agency (North Carolina) **3.** The Sickle Cell Foundation, Inc Central Alabama (Alabama) **4.** SCDAA – Miami-Dade County Chapter, Inc. (Florida) **5.** SCDAA – St. Petersburg Chapter, Inc. (Florida) **6.** Sickle Cell Foundation, Inc. – Tallahassee (Florida) **7.** Sickle Cell Foundation of Kentuckiana (Kentucky) **8.** Sickle Cell Foundation of Georgia, Inc. (Georgia) **9.** Huisman Sickle Cell Foundation of Augusta, Inc. (Georgia) **10.** James R. Clark Memorial Sickle Cell Foundation (Columbia, South Carolina)
NORTHEAST 	Johns Hopkins University	CT, DE, District of Columbia, MA, MD, ME, NH, NJ, NY, PA, Puerto Rico, RI, USVI, VA, VT, WV	**1.** CT – University of Connecticut **2.** DE – Tova Health **3.** District of Columbia – Howard University **4.** MA – Boston Medical Center **5.** MD – Johns Hopkins University **6.** ME – Maine Children's Cancer Program **7.** NH – Dartmouth-Hitchcock Medical Center **8.** NJ – Newark Beth Israel Medical Center **9.** NY – Jacobi Medical Center **10.** NY – (2nd lead) Columbia University Medical Center **11.** PA – Children's Hospital of Philadelphia **12.** Puerto Rico – Universidad de Puerto Rico **13.** RI – Rhode Island Hospital **14.** US Virgin Islands – Virgin Islands Oncology and Hematology **15.** VA – Virginia Commonwealth University **16.** VT – University of Vermont Children's Hospital **17.** WV – Charleston Area Medical Center	**1.** Citizens for Quality Sickle Cell Care* (Connecticut) **2.** Sickle Cell Association of Delaware (Delaware) **3.** William E. Proudford Sickle Cell Fund, Inc. (Delaware) **4.** Faces of Our Children (District of Columbia) **5.** Sickle Cell Association of the National Capital Area, Inc. (District of Columbia) **6.** Armstead-Barnhill Foundation for Sickle Cell Anemia (Maryland) **7.** Association for the Prevention of Sickle Cell Anemia Harford and Cecil Counties and the Eastern Shore* (Maryland) **8.** Christopher Gipson Sickle Cell Moyamoya Foundation (Maryland) **9.** Maryland Sickle Cell Disease Association* (Maryland) **10.** William E. Proudford Sickle Cell Fund, Inc. (Maryland) **11.** Greater Boston Sickle Cell Disease Association* (Massachusetts) **12.** Sickle Cell Association of New Jersey, Inc.* (New Jersey) **13.** Candice Sickle Cell Fund, Inc. (New York) **14.** Queens Sickle Cell Advocacy Network* (New York) **15.** Sickle Cell Awareness Foundation Corp International (New York) **16.** Sickle Cell/Thalassemia Patients Network* (New York) **17.** Children's Sickle Cell Foundation, Inc.* (Pennsylvania) **18.** SCDAA – Philadelphia/Delaware Valley Chapter* (Pennsylvania) **19.** South Central Pennsylvania Sickle Cell Council* (Pennsylvania) **20.** Anemia Falciforme Sickle Cell Disease en Puerto Rico (Puerto Rico) **21.** Life and Family Foundation of Virginia (Henrico, Virginia) **22.** Sickle Cell Association, Inc.* (Virginia) *Chapters of the SCDAA

Data Methodology

Establishment of the Program Measurement System

Data measurements focused on collecting information related to the primary purpose of the Program:

- To establish regional networks and provide leadership and support for regional and statewide activities that will develop and establish systemic mechanisms to improve the prevention and treatment of Sickle Cell Disease by:
    - Increasing the number of providers treating individuals with sickle cell disease using the National Heart, Lung and Blood Institute (NHLBI) Evidence-Based Management of Sickle Cell Disease Expert Panel Report (National Heart Lung and Blood Institute, 2014);
    - Using telementoring, telemedicine and other provider support strategies to increase the number of providers administering evidence-based sickle cell care; and
    - Developing and implementing strategies to improve access to quality care with emphasis on individual and family engagement/partnership; adolescent transitions to adult life; and care in a medical home.

Data Collection Requirements

Per the FOA, the RCCs collected data in the following areas:

- Number of people living with SCD who were served by the Program in the previous year
- Number of eligible patients receiving a hydroxyurea (HU) prescription at least twice in the past year who were seen by participating providers (eligibility for HU as determined by the NHLBI Expert Panel Report)
- Number of people living with SCD seen at participating institutions who have documented recommended pneumococcal vaccinations at least annually (refer to NHLBI Expert Panel Report for pneumococcal vaccination recommendations in SCD)
- Number of eligible patients seen at participating institutions who have documented Transcranial Doppler Ultrasounds (TCDs) at least annually (eligibility for TCD as determined by the NHLBI Expert Panel Report)
- Number of eligible adolescents with SCD seen at participating institutions who have a documented transition plan
- Number of providers in the region who participate in telementoring/telemedicine
- Number of providers by state who participate in telementoring/telemedicine

Develop Data Collection Strategies

RCCs submitted quarterly data from sites to the NCC. Each region was required to work with state-level partners to collect standardized data elements. Each region developed a strategy to implement and maintain DUAs and centralized Internal Review Board (IRB) approvals for data submission for the region. As applicable, RCCs were required to have approved IRB protocols from each of the funded state-level partners within one year of the start date of the award. Regions could include data collected from unfunded state partners within the region to the extent possible. Regions were required to maintain and update (as needed) the data strategy and DUAs to allow for additional measures. Data were to include the ability to report on the number of individuals served by the Program in the previous year.

Data Collection Streams

Data and measurement activities were supported via two primary data collection streams:

1. Provider Survey for Performance Measurement (PSPM) conducted annually through a provider survey
2. Clinical Quality Improvement Measures (CQIM), collected by each RCC and their participating sites quarterly. Each measure set is described in Table 3 and shows the program objectives measured. The schedule of data collection is shown in Figure 2.

All survey instruments were approved by the Office of Management and Budget.

The Report to Congress will use the term Provider Survey for Performance Measurement (PSPM) throughout the text. Some data reports submitted to HRSA during the course of the Program used the term Performance Measurement (PM) when referring to this survey.

The Report Congress will use the term Clinical Quality Improvement Measures (CQIMs) for the quarterly review of medical records. Some data reports submitted to HRSA during the course of the Program used the term Quality Improvement (QI) when referring to these data.

Table 3. SCDTDRCP Quantitative Measure Sets

PROVIDER SURVEY FOR PERFORMANCE MEASURES (PSPM)				
COLLECTED ANNUALLY (2019, 2020)				
ELECTRONIC OR PAPER SURVEY SENT TO PROVIDERS				
1. Number of providers in the SCDTDRCP Network	**2.** Number of patients seen by an SCDTDRCP network provider in the past year	**3.** Number of providers in the SCDTDRCP Network participating in telementoring for SCD in the past year	**4.** Number of SCDTDRCP providers who reported feeling comfortable treating people living with SCD	**5.** Number of SCDTDRCP providers who saw at least one patient in the past year that prescribed hydroxyurea (HU)
CLINICAL QUALITY IMPROVEMENT MEASURES (CQIM)				
COLLECTED QUARTERLY*+ (2019-2021)				
EMR DATA PULL OR MANUAL CHART REVIEW COMPLETED				
1. Hydroxyurea (HU) Use: measured by prescription rates (REQUIRED)	**2.** Transcranial Doppler (TCD) screening	**3.** Immunizations	**4.** Transitions in Care	**5.** Project ECHO® (provider-to-provider telementoring)

Note: All RCCs were required to collect HU use data. They also were required to select at least one additional measure.

* Four of the five RCCs collected data quarterly. The fifth RCC collected data every six months.
+All sites that initiated data collection may not have submitted data every quarter of the Program

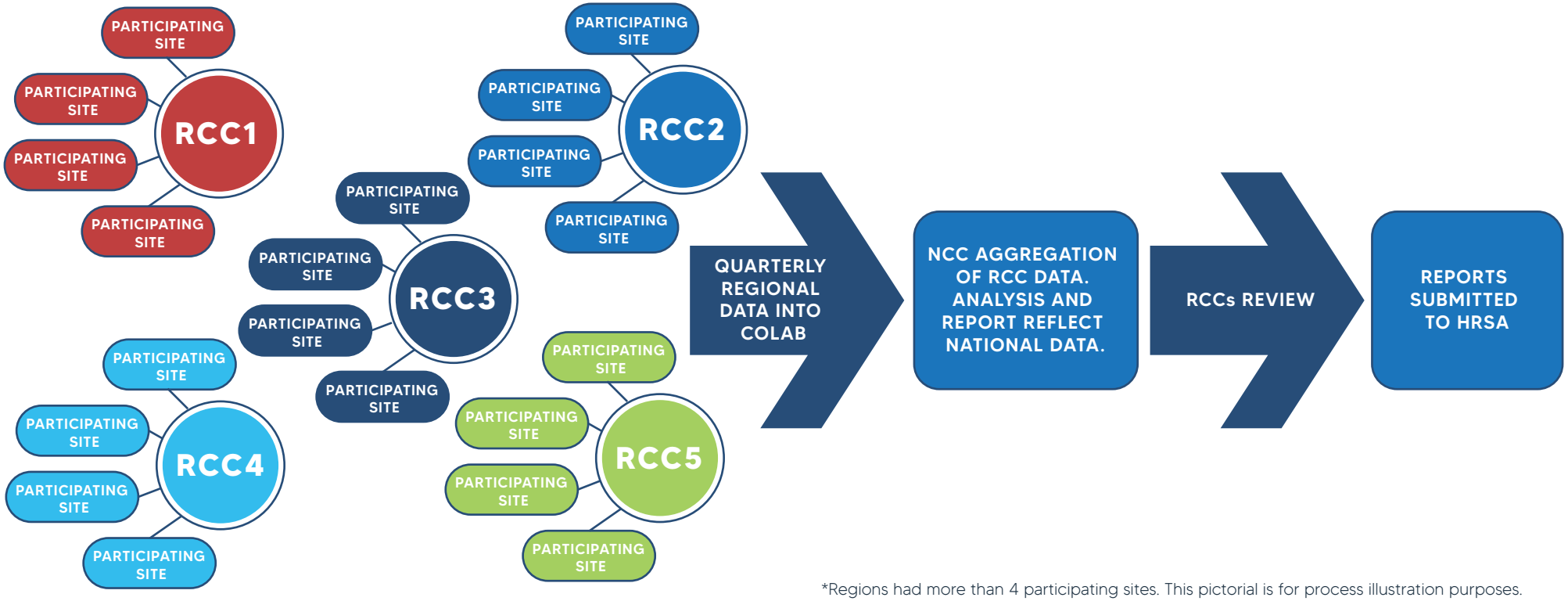
This table outlines the Program objectives and the measures intended to capture applicable information.

Table 4. SCDTDRCP Data Measurement Alignment

THE PROGRAM GOAL	PROGRAM OBJECTIVES BY 2021	MCHB/HRSA EXPECTATIONS OF DATA COLLECTION	PSPMs	CQIMs
1. Increase coordination and service delivery of guideline-based care for individuals living with SCD	1. Each region and at least five funded states participating in the award will have a Sickle Cell Action Plan to increase access to evidence-based care for all individuals with sickle cell disease. 2. Increase by 10 percent from baseline the number of eligible individuals with sickle cell disease receiving an HU prescription at least twice in the past year among patients seen by participating providers. 3. Increase by 10 percent from baseline the number of individuals with sickle cell disease seen at participating institutions that have documented recommended pneumococcal vaccinations at least annually. 4. Increase by 10 percent from baseline the number of eligible individuals with sickle cell disease seen at participating institutions that have documented Transcranial Doppler Ultrasounds (TCDs) at least annually. 5. Increase by 10 percent from baseline the number of eligible adolescents with sickle cell disease seen at participating institutions that have a documented transition plan.	• Number of states in each region that have the required Sickle Cell Action Plans • Number of eligible individuals with sickle cell disease receiving an HU prescription at least twice in the past year that were seen by participating providers. (Eligibility for HU as determined by the NHLBI Expert Panel Report) • Number of individuals with SCD seen at participating institutions that have documented recommended pneumococcal vaccinations at least annually. (Refer to NHLBI Expert Panel Report for pneumococcal vaccination recommendations in sickle cell disease) • Number of eligible individuals with sickle cell disease seen at participating institutions that have documented TCDs at least annually (eligibility for TCD as determined by the NHLBI Expert Panel Report) • Number of eligible adolescents with sickle cell disease seen at participating institutions that have a documented transition plan	**Program Objective 2:** Number of Program providers who saw at least one SCD patient in the past year and also prescribed hydroxyurea • By total • By specialty/ subspecialty • By provider location (Rural/Urban) **Program Objective 2:** Number of patients who were seen by a Program network provider that had an HU prescription in the past year	**Program Objectives 1 & 2:** • HU Use: measured by prescription rates (REQUIRED) • Use of other disease-modifying therapies **Program Objectives 1 & 3:** • Percentage of patients seen at participating institutions that have documented recommended pneumococcal vaccinations at least annually • Percentage of patients who are up to date with vaccinations **Program Objectives 1 & 4:** • Completion of Transcranial Doppler of patients ages 2-16 within the last 15 months **Program Objectives 1 & 5:** • Number of patients that have a documented education discussion/ appointment about transition from Pediatric to Adult Care
2. Increase Access to Quality Care	1. Each recipient will have the ability to report on the number of individuals with sickle cell disease served by the program in the past year. 2. Increase by 10 percent from baseline the number of providers treating individuals with sickle cell disease in each state using the NHLBI Expert Panel Report recommended treatments and prevention.	• Number of individuals with sickle cell disease served by the program in the previous year	**Program Objective 1:** Number of SCD patients seen by a Program network provider in the past year • By total • By specialty/subspecialty • By provider location (Rural/Urban) **Program Objective 2:** Number of providers in the Program Network • By primary practice location type • By specialty/ subspecialty • By provider location (Rural/Urban)	

THE PROGRAM GOAL	PROGRAM OBJECTIVES	MCHB/HRSA EXPECTATIONS OF DATA COLLECTION	PSPMs	CQIMs
3. Increase coordination and service delivery of guideline-based care for individuals living with SCD	1. Increase by 10 percent from baseline the total number of providers, including primary care providers, participating in telementoring and telemedicine activities.	• Number of providers in the region who participate in telementoring/telemedicine • Number of providers by state who participate in telementoring/telemedicine • Number of providers (collated by region and state) who participate in provider support strategies (list out the strategies)	**Program Objective 1:** Number of providers in the Program Network participating in telementoring for SCD in the past year • By specialty/ subspecialty • By primary practice location (Rural/Urban **Program Objective 1:** Number of Program providers that report feeling comfortable treating SCD patients • By provider type • By practice location (Rural/Urban)	• Number of providers participating in Project ECHO® (provider to provider telementoring) or telementoring calls

Figure 2. CQIM Data Collection Process*



*Regions had more than 4 participating sites. This pictorial is for process illustration purposes.

Sources of Qualitative Data

In addition to quantitative data, there were two additional data sources that the NCC used to gather qualitative information about the RCCs and their participating sites' work:

1. Instead of completing in-person site visits, the NCC conducted key informant interviews via Zoom. A qualitative analysis with RCC leads and three local providers from each region to capture salient themes was completed (May-December 2020). RCC leads recommended the three local providers to be interviewed.

2. The NCC collected PowerPoint slides from RCC regional and annual RCC/HRSA grantee meetings conducted during the duration of this funding. These slides were reviewed for additional information about SCD-related programs, research, and clinical improvements, with select examples included in this report.

The Collaboratory for Data Collection (NICHQ's CoLab)

RCCs submitted regional aggregated data into NICHQ's Collaboratory, known as the "CoLab" data portal. This password-protected system was the repository for the quarterly CQIM measures and the annual PSPM data from grantees. NICHQ's CoLab operates within a secure boundary defining a comprehensive set of physical, infrastructure, and application-level controls that protect data submission, storage, and transmission. Only pre-authorized RCC staff had access to the system, and during each session's authentication, users were prompted to accept a data use agreement stating that all data will remain the property of the grantee and HRSA. All user accounts were carefully managed by the NCC and the NCC adhered to an extensive inventory of HRSA policies and procedures to ensure the integrity of the system and the data. The NCC maintained all HRSA Office of Information Technology, Security Operations Center, and Office of Information Security & Privacy regulations.

Developing, Refining and Maintaining the Manual of Procedures

Building on prior Program experience, the NCC, with input from the RCCs, HRSA/MCHB, and the OSC, documented the data collection processes in the MOP. The MOP includes background Program information, data dictionaries for the CQIM and PSPM questions, and additional data collection guidance documents.

Prior to data collection, the NCC reviewed, troubleshot, and resolved issues related to the data dictionary (October-December 2018). RCCs were given the MOP for final comment and review in early 2019. Based on the MOP, NICHQ's CoLab was programmed to accept data outlined in the data dictionary.

The MOP included the following information about the Program data collection, entry, and analysis:

- Orientation to NICHQ's CoLab
- Project-specific data collection and entry instructions
- Program measurement and data overview
- Timeline for data collection
- Data Collection Protocol, including requirements for data submission, programming for PSPM and CQIM data collection forms, and content for obtaining measures
- User guide for how to enter data into CoLab
- Hydroxyurea (HU) listings from the National Drug Code Directory
- Immunization schedules for people living with SCD
- CQIM REDCap Survey Build (developed and shared by Pacific RCC as a sample)
- Sample regional aggregation filter logic (developed and shared by Pacific RCC)
- PSPM questions
- PSPM RCC reminder prompts

The MOP was updated regularly to reflect relevant decisions and clarifications regarding data collection. When questions or clarifications arose, the NCC team facilitated conversations among RCCs, HRSA/MCHB, and the OSC to come to a resolution. All MOP changes were discussed during group data management working meetings as well as one-on-one calls with the RCC teams. Final decisions were documented in the MOP.

Table 5. Changes Made to the MOP During the Program

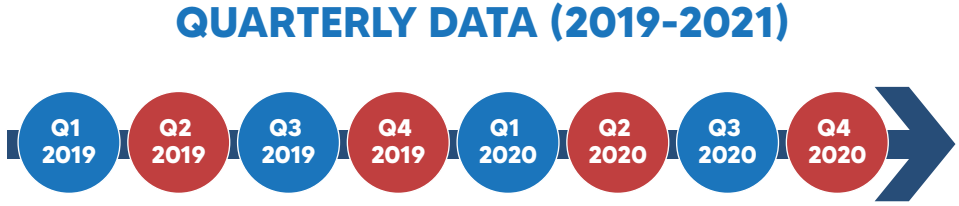
PSPM- OR CQIM-RELATED	CHANGES
CQIM: Measure 4	Clarification of how to calculate denominators for immunizations
CQIM: Measure 4	Clarification of "up-to-date" status
CQIM: Measure 4	Added Immunocompromised status with immunizations
CQIM Measures (overall)	Added number of sites reporting
CQIM: Measure 4	Added status of Hib vaccination series
PSPM	Correction of known incorrect responses
PSPM	Added multiple responses from same provider
PSPM	Non-numeric responses reported for zip codes
PSPM	Added ranges reported for patients seen, HU use, etc.
CQIM: All denominators	Added patient panel definition
CQIM: Immunizations	Added accounting for Flu Shots by Flu Season and Quarter
CQIM	Edits to Q2 QI 2021 timeline – with HRSA approval, shortened this quarter to accommodate earlier fiscal completion of RCCs than the NCC
PSPM	Changes to PSPM survey timeline for fielding of second survey due to COVID-19 impact
CQIM/ PSPM	Updated Gantt chart to reflect 2020/2021 timeline edits

Standardizing Data Collection from Grantees

The NCC worked with the RCCs and HRSA/MCHB to manage consistent, standardized data collection. The NCC organized multiple opportunities for communication across all Program stakeholders. Tailored technical assistance was provided to RCCs during individual monthly meetings. These meetings addressed regional variables that impacted an RCC's data collection. During these meetings, the NCC relayed communication from HRSA/MCHB as well as reviewed relevant data collection strategies and decisions. Group technical assistance was provided during monthly DMWG meetings. These meetings ensured communication continuity regarding the Program purpose and goals while underscoring unified decisions related to the direction of data collection and measurement. These meetings also served as a forum to facilitate discussions for common processes and problem resolution.

Data Collection Time Points

Figure 3. Quarterly CQIM Data Submission Timeline



Office of Management and Budget (OMB)

Data collected for this HRSA/MCHB program was subject to all applicable federal laws, which limit the burden that can be placed on grantees. To assess the burden of data collection, the OMB required submission of an information collection request package for the Program PSPM and CQIM. HRSA developed and submitted the information package in coordination with the NCC. To inform the OMB information collection package, various methods of obtaining data were considered to assess the burden of data collection. The 60-day Federal Register Notice (FRN) seeking public comment on the information collection request was published in January 2020 and a subsequent 30-day FRN was published in July 2020. No comments were received regarding the survey instruments during either period. Full approval was received in September 2020, allowing formal data collection to begin. See OMB website for full information.

Overview of Data Sources

Provider Survey for Performance Measurement

RCCs conducted an annual assessment (May 2019 and September 2020) of the PSPM via a survey of network providers. The PSPM data collection activity was to assess key areas of SCD care. The PSPM survey was developed by HRSA and refined with RCCs and NCC input. RCCs disseminated the survey within their regions. Survey distribution was primarily completed using the REDCap (Research Electronic Data Capture) program, but paper surveys were available as requested.

Assessment Areas for the PSPM

- Network size
- Characteristics of patients with SCD seen in past year
- Telementoring activities
- Comfort level of network providers with SCD care
- Hydroxyurea use

RCCs aggregated regional responses and submitted to the NCC via NICHQ's CoLab. The NCC then aggregated regional data and created a report.

Response rates from the two provider surveys conducted are listed in Table 6 and Table 7. Results and discussion of the PSPM are located in the reports produced for these data collection points in Appendix B.

Table 6. Response Rate for 2019 PSPM Survey by Region

REGION	NUMBER OF SURVEYS SENT	NUMBER OF SURVEYS COLLECTED	% RESPONSE RATE
Overall	1854	516	27.8%
Heartland/Southwest	163	51	31.3%
Midwest	219	79	36.1%
Northeast	292	34	11.6%
Pacific	79	38	48.1%
Southeast	1101	314	29.0%

Table 7. Response Rate for 2020 PSPM Survey by Region

REGION	NUMBER OF SURVEYS SENT	NUMBER OF SURVEYS COLLECTED	% RESPONSE RATE
Overall	1220	306	25.1%
Heartland/Southwest	241	73	30.2%
Midwest	160	50	31.3%
Northeast	378	61	16.1%
Pacific	91	43	47.3%
Southeast	350	79	22.6%

PSPM Survey Network Definitions

For the first PSPM survey, RCCs individually determined their "network" of providers – the people who were sent the survey (Table 8). For the second PSPM survey, it was decided to have a standard definition of "network" that would be used by all RCCs so that a uniform comparison between regions could be conducted. After discussions with the NCC and RCCs, HRSA/MCHB developed "a network" guidance document. This guidance document was used to determine which providers should be sent the second survey.

The NCC shared the final document (Table 9) with the RCCs at the March 2020 DMWG meeting. Because the pool of potential respondents was not consistent across the two time points, survey responses between timepoints should not be directly compared. Rather, they should be considered as two distinct cross sectional data points. Should the network definition stay the same in the future, the second survey can be used as a baseline assessment.

Table 8. Regional Definitions of Provider Network Used for 2019 PSPM Survey Distribution

REGION	REGIONAL NETWORK DEFINITION USED FOR PSPM SURVEY #1
Heartland/ Southwest	The Heartland/Southwest identified contacts from local provider list-servs (e.g., hematology/oncology providers, emergency department providers) and locally developed lists of contacts that local sites felt would be most likely to respond. All providers were considered to be able to regularly treat patients with SCD and to prescribe HU if indicated. The Heartland/Southwest RCC defined their network of providers as meeting one or more of the following criteria. All providers who: • Regularly treated SCD and prescribed HU • Were selected by the local site leads • ECHO participants, excluding contacts external to the region
Midwest	The Midwest RCC defined its network as prescribing providers within the Midwest region who met one or more of the following criteria: • Attended at least one of the region's ECHO® sessions or educational programs given by the state leads • Prescribing providers as selected by site leads, including those that were partnering with the state lead
Northeast	The Northeast RCC defined its network as: • Any licensed prescriber working in the region's states/territories/districts in the Northeast region • Note, this region also included any licensed prescriber that had participated in at least one of their hosted ECHO sessions (providers may have been outside of the Northeast geographical region)
Pacific	The Pacific RCC defined its provider network as: • Providers who attended their Project ECHO® sessions, except residents and providers who do not prescribe HU (e.g., social workers, psychologists) • Select others who the site leads know provide clinical care to the population with SCD in the region
Southeast	The Southeast RCC allowed each local site/state lead to define individual strategy outreach. There were multiple ways the RCC defined and collected their list of providers, including: • Review of listservs • Review of state medical society membership • Review of rosters of local provider networks that may treat patients with SCD in emergency departments, community health centers, local hospitals, and medical centers of SCD excellence • Project ECHO® participants • Outreach to known providers of SCD care in a state • Review of local ASH referral sites • Review of contact rosters accrued from site visits, regional conferences, and local SCD meetings within the region

Table 9. Standardized Definitions of Provider Network Used for 2019 PSPM Survey Distribution

	STANDARDIZED PROVIDER NETWORK DEFINITION USED FOR PSPM SURVEY #2
Definition	The Provider Survey should be sent to "SCDTDRCP Providers" defined as those providers for whom SCDTDRCP **funding** could conceivably **lead to causing changes** in whether they see SCD patients, their comfort seeing SCD patients, or prescribing of disease-modifying therapies, particularly hydroxyurea. In other words, we want to include providers (of all specialty) for whom we can identify meaningful contacts with the Program. Based on your list of touchpoints, "SCDTDRCP" providers include those that: • Participated in SCDTDRCP-sponsored ECHO sessions • Are SCDTDRCP state leads • Attended SCDTDRCP CME/MOC* presentations or grand rounds • Have clinical care discussions and/or care coordination with SCDTDRCP state leads • Participated in SCDTDRCP-sponsored intensive education (e.g., boot camp) • Participated in SCDTDRCP-sponsored provider summit and trainings • Participated in SCDTDRCP-led QI projects • Attended SCDTDRCP-led resident teaching Note: If a provider qualifies based on this list, they MUST be included in the survey sample (We recognize that if you do not have an email address or other mechanism to contact the provider, they will not be included in the sample). **Continuing Medical Education/Maintenance of Certification*

Effects of COVID-19 on the PSPM Data Collection

Originally, three annual PSPM surveys were planned. The launch of the Year 2 survey was originally slated to begin May 1, 2020. However, the second annual survey was initiated later than planned due to the COVID-19 pandemic.

In January 2020, the United States declared a public health emergency related to the COVID-19 pandemic. Many of the Program providers at the local and regional levels were engaged in either front line care or planning and development of processes to respond to emerging needs. By April 2020, it was clear to HRSA that fielding the provider survey in May 2020 as originally planned was not a reasonable ask of RCCs or potential respondents. HRSA made the final decision on April 7, 2020, to postpone the survey to a later date. HRSA asked the NCC for recommendations about next steps and possible options. Following the NCC's recommendations, HRSA decided to field only one additional provider survey (vs. a planned two additional rounds of fielding). The final survey was launched in September 2020.

Clinical Quality Improvement Measures Data Collection

Clinical Quality Improvement Measures (CQIM)

RCCs collected CQIM data quarterly. See Figure 2 for the Data Collection Process. RCCs were required to collect and report information about hydroxyurea (HU) and at least one additional CQIM of their choice.

RCCs coordinated the work of the local sites within their regions to collect the data. The CQIM measures collected were intended to reflect population-level data on key measures related to the Program (Figure 4). Data for these measures were obtained from Electronic Health Records (EHRs) or manual medical record review at the participating sites. RCCs aggregated CQIM measure data quarterly and submitted corresponding numerators and denominators to NICHQ's CoLab. Four of the five RCCs collected data on a quarterly basis. One RCC provided data on a six-month schedule (reporting for Q2, Q4). All data were re-run to include any revisions on a six-month basis (at Q2 and Q4). The Q1 2019 through Q4 2020 CQIM reports can be found in Appendix B.

Barriers to Data Collection

Only data collection of HU prescription rates were required. RCCs were asked to select at least one additional area to capture data.

Because of this, there was variation across regions regarding the number of sites who submitted data for additional measures. Staffing limitations and other clinic site factors also impacted consistent data submission. Fluctuations in the denominator reflect some of these variations.

There were challenges to collecting each of the data measures, however, collecting immunizations was the most problematic. Up to date status requires both immunization delivery and documentation of the delivery. As noted in the report, Program teams encountered several challenges collecting these data. For example, depending on the state, some hematologists and oncologists can give all immunizations, while others cannot and must rely on the pediatrician or PCP to administer. Access to these reports can be problematic, thus complete records may not exist with a single provider or healthcare system. Providers caring for patients who live in another state faced a unique concern. These providers may not have access to a neighboring state's database, with a faxed request the only way to obtain official documentation. The staffing was not always available for these types of tasks. With disparate ways of getting immunizations, the Program found that information was not uniformly getting transferred, fracturing record keeping and making collection inconsistent.

Definition of transitional care was not uniform across either regions or sites. Additionally, COVID-19 brought on other data collection barriers listed in this report.

Interpretation of the Data

The following should be considered:

- The data were collected through convenience sampling
- Measure denominators fluctuated over time due to variation in sites reporting each quarter
- The COVID-19 pandemic coincided with the 2020 data collection period

Given these factors, definitive assessment of improvements in program objectives across time points is not possible and findings are not fully generalizable.

Data were collected for 2021 but are not included in this report. Please contact Alisha Keehn (akeehn@hrsa.gov) at HRSA to obtain 2021 reports.

Changes to the Clinical Quality Improvement Measures

During the Program, there were minor changes and clarifications to CQIM measures. For example, one change was related to CQIM Measure 3: immunizations, which posed several collection challenges. Originally, the data dictionary directed that the denominator for all sub-measures under immunizations represent the full population of patients eligible for any immunization being tracked. However, during discussions with RCCs, it was suggested that the denominator be specific to each immunization collected because of substantial limitations in accessing population level data and that providing a denominator of patients who were eligible to receive a particular immunization would be a more accurate measure of the completion of the immunization. The lack of reliable data across EHRs and the variability between state immunization record systems made this data particularly difficult and burdensome to systematically collect. Therefore, after discussion with RCCs and HRSA/MCHB in February 2020, the NCC revised the CQIM Data Dictionary to direct that starting in Q1 2020, immunization data collection be submitted on an intent-to-treat model versus population-level information. The NCC updated the MOP to correct the specifications for the denominator within each immunization measure to reference patients eligible for a specific immunization. Please see the MOP for data dictionary details. Starting in Q1 2020, data reports noted this change in a footnote.

Effects of COVID-19 on the CQIM Data Collection

In March and April 2020, the NCC held conversations with RCC teams during individual monthly check-in calls to learn how the pandemic was impacting programs, local sites, and capacity to collect data. While all sites were in favor of canceling or postponing the planned initiation of the second PSPM in May 2020, all believed they could collect and submit CQIM data as scheduled. The NCC requested, and RCCs agreed, to be in close contact and communicate changes in CQIM data submission. RCCs relayed pertinent information. For example, in Q1 2020, the Midwest and Southeast regions both had one local site that was unable to submit data due to staff furloughs. The RCCs annotated in NICHQ's CoLab that data were missing for relevant measures from the site; the NCC noted the fluctuation of sites submitting data during the course of the Program.

Qualitative Data Collection

To collect a comprehensive picture of the activities of the Program, qualitative data were collected. Data collection was initially planned as face-to-face interviews, but these were conducted virtually due to the COVID-19 pandemic. In May 2020, all five RCC leads were interviewed. During these interviews, each RCC lead was asked to recommend three participating sites for interviews with the NCC. Not all RCC sites were interviewed for this report due to time considerations. Information from the interviews with sites, including select quotes and RCC activity examples, is reflected in greater detail in the report and in Appendix A: RCC Activities. Participating site interviews occurred between July and August 2020. Table 10 provides an overview of the RCC and local site interviews conducted.

Qualitative Process

The NCC created a protocol and conversation guide for RCC interviews and for participating site interviews. Questions related to challenges and successes and specifics regarding their role as an RCC or as a participating site (as applicable). After each interview, the NCC prepared transcripts of the discussion. All RCC interviews were transcribed using Zoom and manually edited. The local site interviews were transcribed by Zoom/manual editing or a professional transcription service. Using the transcription, two NCC staff read each line of the transcript and used a qualitative analysis technique, called inductive coding. That is, after the NCC staff read the interviews, they came up with codes for different topics. From this, a codebook was created for the RCC interviews. For local site interviews, the NCC used the codes that emerged from the RCC codebook and added codes for content that did not arise in the RCC interviews. For both RCC and participating site interviews, the NCC staff independently coded 3-4 interviews and then met to review coding decisions, agreements, and differences. Two members of the code team then independently coded the remaining interviews using the codebook. Once coding was completed, the NCC identified high level themes, categorized exemplar quotes, and summarized content.

Table 10. Local Site Qualitative Interviews

RCC	LOCAL SITE
Heartland/Southwest	Truman Medical Center, Kansas City, MO
Heartland/Southwest	Baylor College of Medicine & Texas Children's Hospital, Houston, TX
Heartland/Southwest	University of Arkansas for Medical Sciences, Little Rock, AR
Midwest	OSF Healthcare Children's Hospital, IL
Midwest	Children's Hospital of MI, Detroit SCDAA, MI Chapter
Midwest	Children's Hospital, MN Vascular Anomalies Center
Northeast	Jacobi Medical Center
Northeast	Charleston Area Medical Center, WV
Northeast	William E. Proudford Fund, Inc.
Pacific	Sickle Cell Disease Foundation of California
Pacific	Oregon Health and Science University
Pacific	Arizona, Phoenix Children's Hospital Children's East Valley Specialty
Southeast	Prisma Health -Upstate, Greenville SC
Southeast	Medical College of Georgia, Augusta, GA
Southeast	University of Louisville School of Medicine Affiliated with Norton Children's Cancer Institute

References

National Heart Lung and Blood Institute. (2014). Evidence-based management of sickle cell disease: Expert panel report, 2014. https://www.nhlbi.nih.gov/sites/default/files/media/docs/sickle-cell-disease-report%20020816_0.pdf

SICKLE CELL DISEASE
TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Model Protocol: Strategies and Resources for Replicating the Program

NICHQ
National Institute for Children's Health Quality

SEPTEMBER 2021

Introduction

Model Protocol

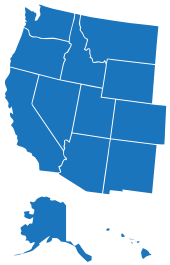
Introduction

This document contains information for the Model Protocol for the Sickle Cell Treatment Demonstration Collaboratives Program (SCDTDRCP). Between 2017 and 2021, Health Resources Services Administration (HRSA) funded this Program as part of the Sickle Cell Disease and Other Heritable Blood Disorders Research, Surveillance, Prevention, and Treatment Act of 2018, 42 U.S.C. § 300b-5 (2018). The Program description and findings can be found in the 2021 Congressional Report.

NOTE:
Throughout this report, the Sickle Cell Disease Treatment Demonstration Regional Collaboratives Program is referred to as "SCDTDRCP" or "the Program."

The purpose of this Model Protocol is to offer providers, community-based organizations (CBOs), public health agencies, academic institutions, healthcare organizations, policy makers, and others guidance on promising practices and strategies developed when providing care for people living with SCD. This tool was developed to support the spread and implementation of clinical guidelines and development of systems of care that can collectively improve the health and quality of life for people living with SCD. These resources and strategies have been compiled from the current RCCs and their state partners. They have been reviewed and identified as effective strategies by leaders advancing SCD care at local comprehensive sickle cell centers, regional centers of collaborative learning, and experts from across the nation. The recommended strategies work best within collaborative teams via collective feedback and through tailoring by key stakeholders that comprise the system.

The Program network was divided into five Regional Coordinating Centers (RCCs) representing 50 states and territories:



1. Pacific Sickle Cell Regional Collaborative (PSCRC)

2. Heartland/Southwest Sickle Cell Disease Network

3. Sickle Treatment and Outcomes Research in the Midwest (STORM)

4. Sickle Cell Improvement in the Northeast Region through Education (SINERGe)

5. Education and Mentoring to BRing Access to CarE (EMBRACE) in the Southeast

The Program built upon and expanded work begun in the previous cycles of this funding in 2014 and 2017. These initiatives were intended to develop and refine clinical protocols and to identify priority areas of care coordination to ensure that all patients receive the highest quality of care. In 2014, a Compendium of Tools and Resources was created that was designed to help spread essential tools and resources developed and tested over the course of the Program. This Compendium was updated in 2017 and again in 2018. A newly released 2021 version is part of the 2021 Report to Congress. The Compendium holds key tools and resources for providers and teams working with patients and families. It is designed to facilitate the provision of high-quality care for children and adults living with SCD and sickle cell trait (SCT) and select resources are referenced in this document.

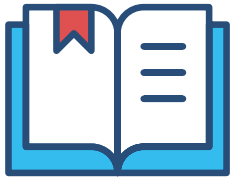
The five RCCs used a diverse set of Quality Improvement (QI) tools to test, implement, and spread effective, evidence-based clinical protocols and system changes to address the aims of this Program. While these change ideas have been tested and implemented across communities and regions, it is recommended that QI principles be employed when implementing these change ideas in new settings (Institute for Healthcare Improvement, 2020). Using QI principles to apply and test these ideas in a new environment can better ensure that local care needs are met.

Organization of the Model Protocol

This Model Protocol includes the resources, strategies, and best practices organized into four sections reflecting broad areas of Program work:

1. Increasing Access to Quality Care

2. Increasing Delivery of Hydroxyurea, Immunizations, and Transcranial Doppler



3. Increasing Provider Knowledge Through Education

4. Increasing Use of Technology That Supports Healthcare Delivery

Each of these sections includes an overview narrative, followed by a table that lists domains and rationale for activities along with resources.

SICKLE CELL DISEASE TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Strategies and Resources to Increase Access to Quality Care

Model Protocol

Strategies and Resources to Increase Access to Quality Care

SCD is a serious genetic condition that, while rare, can have a significant impact on affected people and their families. Those living with SCD can have acute pain episodes and are at high risk for complications, such as infection, acute chest syndrome, and stroke. These complications can have a severe impact on both the quality of life and overall lifespan for people living with SCD. Because of this impact, receiving high quality care is important. However, there are wide-ranging barriers to accessing quality care for people living with SCD.

Comprehensive SCD care teams have been established to provide coordinated care that meets current clinical guidelines. However, these teams or centers may not be accessible to people who reside in areas, such as rural communities far away from large academic health systems, preventing them from receiving new and innovative technology and enhanced treatment options that could help them live longer. Particularly, adults living with SCD may experience significant difficulties accessing quality primary and specialty care. The reasons for this disparity are numerous and include a lack of knowledgeable providers able to care for people living with SCD. Primary care providers frequently have a limited number of people living with SCD on their clinical panels, which diminishes opportunities to enhance expertise and stay up-to-date with contemporary guidelines and recommendations related to caring for people living with SCD.

INCREASING ACCESS TO CARE

RCCs used a multidimensional approach to increase quality care for people with SCD, including:

- Enhancing community partnerships
- Expanding program and provider availability
- Improving transition from pediatric to adult care
- Addressing financing

Enhancing Community Partnerships

RCCs created integrated care networks by partnering with key stakeholders and communities to increase the reach of clinical care sites within communities and connect with hard-to-reach populations. This work was supported by the development of close partnerships with CBOs that support children, adolescents, adults, and families living with SCD in their homes and communities. In addition, providers and CBOs worked together to utilize community health workers to identify and connect individuals to both primary and specialty care and address social determinants of health. These strategies helped create a bridge to additional supports for families affected by SCD and to extend the reach of clinical sites within the communities they serve.

Expanding Program and Provider Availability

In rural areas where geographic barriers challenge care access, state and regional partners coordinated transportation; set up satellite centers of care; collaborated with advanced practice professionals (APPs); and leveraged telehealth to reach rural patients to ensure they had routine SCD care. RCCs increased the use of telementoring via regional and local Project Extension for Community Healthcare Outcomes (ECHO®) sessions to bolster provider-to-provider education among providers (including specialists, PCPs, and APPs (both urban and rural providers). Project ECHO® was developed in 2002 by Sanjeev Arora, MD, at the University of New Mexico and was a key focus for the Program teams. Project ECHO® is an innovative telementoring program designed to create virtual communities of learners by bringing together healthcare providers and subject matter experts using videoconferencing to provide brief lecture presentations and case-based learning, fostering an "all learn, all teach" approach. Participants are engaged in the bidirectional virtual knowledge network by sharing clinical challenges and learning from peers and specialty experts across the country. Project ECHO® has been globally recognized as a successful tool to improve patient care outcomes.

Improving Transitional Pediatric to Adult Care

The life expectancy of people living with SCD has increased. However, the period of transition from pediatric to adult medicine is identified as a time of increased risk of morbidity and mortality. Thus, a robust transitional process during this time is essential. In many regions, provider organizations are central to the creation of systems-level changes and resources to track and facilitate transitions from pediatric to adult care. In addition, CBOs are increasingly helping to develop adolescents' life skills and capacity to self-manage aspects of their condition with specialized training curricula. Partnership between CBOs and healthcare organizations is important for reaching patients where they are, particularly at crucial time periods, such as young adulthood when the transfer from pediatric to adult healthcare systems occurs.

LIFE EXPECTANCY

The life expectancy of people living with SCD has increased. However, the period of transition from pediatric to adult medicine is identified as a time of increased risk of morbidity and mortality.

Addressing Financing

Along with established strategies to increase the number of providers and institutions delivering SCD care, RCCs discussed the need to address healthcare financing policies to allow for adequate reimbursement for health services. SCD treatment programs and other stakeholders are creating coalitions to work with local and state policymakers along with payer groups to educate about the need for appropriate reimbursement and the importance of having payment arrangements in place to support providers' ability to improve access to quality care. For example, reimbursement policies that cover coordination between specialists and primary care providers can facilitate and support seamless care. Funding made available for telehealth needs can result in better disease management and quality of life for people living with SCD.

Learning From the Program Work: The Following are Suggested Approaches to Increasing Access to Care for People Living with SCD

- Facilitate connections between CBOs and healthcare providers to ensure maximized outreach and healthcare access for people living with SCD and their families (e.g., grow and expand mutual partnerships with local organizations)
- Identify and connect people living with SCD, especially young adults, with providers and systems of care that support self-management, care coordination, and care transitions, such as transferring from pediatric to adult care
- Reach out to and provide educational opportunities for primary care providers, specialists, and APPs to enhance awareness about SCD and outline appropriate referral processes to ensure access to specialty care
- Expand physical and virtual access to care through the development of new programs or satellite locations and leveraging telehealth technologies

The following table provides suggested strategies and related rationales and resources for increasing access to care for people living with SCD.

Table 1. Strategies and Resources to Increase Access to Quality Care

STRATEGY	RATIONALE	RESOURCES

Creation of Sickle Cell Disease Programs	All regions support the development of additional sites for the provision of comprehensive care to ensure availability to all people with SCD. Both quantity and quality of these settings for care matter and several regions have been successful in setting up clinics. See the suggested resources for planning and implementing a comprehensive SCD care team.	• *Creating a New Sickle Cell Clinic: A Case Study: A few dedicated professionals discuss how and why they created a new county clinic in South Los Angeles.*
		• EMBRACE Sickle Cell Disease Network (Setting up a comprehensive SCD clinic)
		• Examples of State plans (Comprehensive List in Compendium of Tools)
		  ○ Pacific RCC State Action Plan
		  ○ Texas State Action Plan

Provider Outreach and Engagement	There are a limited number of providers with expertise in the care of people living with SCD, specifically adults. As people living with SCD live longer with specialized care, providers need to be knowledgeable about this condition and up-to-date recommendations for primary and specialty care for this population.	• Illinois Provider Engagement Survey (contact storm@cchmc.org for information) • *Enhancing Access to Care for Sickle Cell Disease in South Carolina* (webinar) • *Sickle Cell Awareness Guide for Providers* (Midwest RCC)
Enhancement of SCD and SCT Awareness in the Community and Healthcare Institutions	People living with SCD are more likely to access quality care when they are knowledgeable about the care that they need and know where to obtain services. Similarly, communities affected by SCD can better advocate for high-quality, comprehensive care when they are informed about SCD and SCT. Informational materials on SCD and SCT can also be used in the clinical setting by both primary care and providers who specialize in SCD care.	**What You Should Know About Sickle Cell Trait** • English French Spanish (handout) • *Sickle Cell Trait Toolkit* (website) • Midwest RCC: *Cincinnati Children's: Sickle Cell Trait* (website) • Trait Counseling Booklet • SC Trait Explanatory Brochure **Get Screened to Know Your Sickle Cell Status** • English French Spanish (handout) **What You Should Know About Sickle Cell Disease** • English French Spanish (handout) • *5 Facts You Should Know About Sickle Cell Disease* (handout) **What You Should Know About Sickle Cell Disease and Pregnancy** • English French Spanish (handout) • *Pregnancy and Sickle Cell Disease*
Enhancement of Family and School Personnel Awareness About Management of SCD at Home and in Early Childhood and School Settings	It is essential that caregivers and school personnel are knowledgeable about SCD, related symptoms, available treatments, and necessary accommodations for children living with SCD. Resources educating caregivers and teachers about SCD management can help children and adolescents in educational and social settings.	*Tips for Supporting Students with Sickle Cell Disease* (booklet) **Living Well with Sickle Cell Disease: Tips for Healthy Living** • English Spanish (handout) • *5 Tips to Prevent Infection* (handout) • Disease education materials

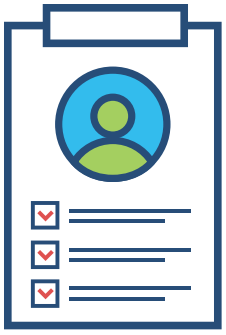
Table 1. (continued) Strategies and Resources to Increase Access to Quality Care

STRATEGY	RATIONALE	RESOURCES
Support for Adolescents Living with SCD Transferring from Pediatric to Adult Care	Many people living with SCD are living into adulthood. Smooth transfer from pediatric to adult care is essential but can be fraught with complications related to changes in insurance, care providers, and care settings. Consequently, it is important to provide education about chronic disease self-management and general life skills to young adults and their families navigating the transition from pediatric to adult care.	• *Nine Steps to Living Well with Sickle Cell Disease in College* (handout) • *Pediatric to Adult Health Care Transition: A Family Toolkit* • Readiness assessment for parents • Readiness assessment for youth • Health Care Transition Timeline for Parents and Caregivers • Health Care Transition Timeline for Youth and Young Adults • Sickle Cell Transition from age 13 to 15 • Sickle Cell Transition from age 16 to 18 • Sickle Cell Transition from age 19 to 21 • Patient Empowerment Toolkit • See Compendium of Tools and Materials for additional resources
Implementation of an SCD-specific Transition Curriculum	Several clinical sites have begun to develop specific, comprehensive curricula that encompass the typical span of the transition period (12-21 years of age). Clinics should first establish a system for transition following the *Got Transition*® six core elements. Curricula should include recommendations of educational content for providers, adolescents and young adults living with SCD, and their families. Use of the entire curricula is recommended so that all topics are covered throughout the transition planning process. Additional areas that should be addressed include guidelines for discussing topics, methodologies of teaching the curricula, and techniques to measure efficacy of the teachings. Curricula can be used as a resource in both the medical and the community setting and are effective in helping organize the work conducted as a partnership.	• A program of transition to adult care for sickle cell disease (ASH Education Program) • Sickle Cell Transition Curriculum • https://www.floridahats.org/

SICKLE CELL DISEASE TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Increased Delivery of Hydroxyurea, Transcranial Doppler, and Immunizations

Model Protocol



Strategies to Increase Hydroxyurea, Immunizations, and Transcranial Doppler

Promising strategies have been employed to increase delivery of Hydroxyurea (HU), Transcranial Doppler (TCD), and immunizations but they have varied greatly from region to region and provider to provider (Wang et al., 2013). The use of evidence-based guidelines, protocols, and shared decision-making tools, both in electronic and print forms, can help facilitate providers' communication with patients and their families about the clinical benefits, side effects, and various considerations related to SCD treatment. Materials also help review applicable long-term consequences of treatments and preventive and screening measures. These resources have been demonstrated to empower patients and families by increasing knowledge of evidence-based treatment options, widening their understanding of risk, and decreasing decisional conflict (Crosby et al., 2015).

Expert Guidelines

The RCCs and participating sites have successfully employed expert guidelines and protocols, such as those published by the American Society of Hematology (ASH) and the National Heart, Lung, and Blood Institute (NHLBI), to facilitate conversation between patients and providers. In addition, several tools have been developed by RCCs and their participating sites, leveraging the NHLBI evidence-based clinical guidelines for use and delivery of HU, TCD, and immunizations in SCD care (National Heart Lung and Blood Institute, 2014). Note: it is anticipated that future iterations of the Model Protocol and Compendium of Tools and Resources will include strategies to enhance the use of newly approved disease-modifying therapies for SCD, such as crizanlizumab (Adakveo) and voxelotor (Oxbryta) (Ali et al., 2020). These therapies are not included in this Model Protocol.

Shared Decision-Making Using Print and Electronic Materials

An expert panel from NHLBI encourages shared decision-making with all patients. Shared decision-making tools can help providers feel more comfortable initiating conversations and ensure that patients are making informed decisions. Materials such as brochures, flyers with infographics, and websites facilitate treatment conversations between physicians and other care team members and patients by outlining key treatment information and considerations. An advantage of these resources is that patients can take them home to read or look at them over time and review with family members. This allows time to develop and bring back questions to discuss with care teams. The Program has successfully created both print and electronic communication materials as ways to support both patients and providers during the care process with the goal of increased uptake of HU, TCD, and immunizations.

Hydroxyurea (HU)

Until July 2017, HU was the only Food and Drug Administration (FDA) approved therapy for SCD (Brawley et al., 2008; Charache et al., 1995; National Heart Lung and Blood Institute, 2014; Yawn et al., 2014). This medication decreases SCD-related complications, such as pain crises, acute chest syndrome, and associated emergency department visits and hospitalizations (Wang et al., 2013). HU can improve the quality of life for patients by reducing the frequency of these complications of SCD (Steinberg et al., 2003; Steinberg et al., 1997; Thornburg et al., 2010). Hydroxyurea has been found to lower the costs associated with care for people living with SCD. While patients who take HU have been found to have higher costs (due to paying for the medicine), these costs are outweighed by the savings from fewer inpatient hospitalizations (Wang et al., 2013). However, many individuals and their families are unaware of the drug or its potential benefits, as their providers do not discuss HU with them, leading to reduced use. Also, some patients who are aware of the therapy may be reluctant to use disease-modifying therapies (Creary et al., 2015; Oyeku et al., 2013). In these cases, providers may be able to address common questions and concerns. Increasing both provider and patient knowledge is key to increasing use of HU. Below are resources for both groups.

Table 2. Strategies and Resources to Increase Delivery of Hydroxyurea

STRATEGY	RATIONALE	RESOURCES
Evidence-Based Guidelines and Protocols for Clinical Practice Support	The use of evidence-based guidelines is recommended as they have been shown to improve the health and healthcare of people living with SCD. Guidelines should be developed using a systematic process, including literature review and grading of the evidence, to address specific clinical questions developed by an expert panel (DeBaun, 2014; Yawn et al., 2014).	*Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014*, NHLBI ASH Pocket Guide: Hydroxyurea and Transfusion Therapy for the Treatment of Sickle Cell Disease *Hydroxyurea for Sickle Cell Disease: Indications, Dosing and Monitoring* (Midwest RCC, Cincinnati Children's Hospital)

Enhancement of Patient Awareness of HU and Shared Decision-Making Tools	Though increasing, HU remains an underutilized treatment for SCD. There are several reasons for underuse of this therapy, including lack of awareness among some providers and patients about its effectiveness and use and concerns related to potential side effects. (Brandow & Panepinto, 2010). To increase acceptance and adherence to HU, patients and families need accurate information so they can discuss their treatment options with their providers. Information can be shared through brochures, flyers, and electronic shared decision-making tools (Thornburg et al., 2010).	*Sickle Cell Disease: Hydroxyurea: What You Need to Know* (NHLBI handout)
		Midwest RCC shared decision-making tools
		• Initial HU visit
		• Recurring HU visits
		• Hydroxyurea White board
		• Hydroxyurea brochure
		Southeast:
		• St. Jude informative hydroxyurea pamphlet
		PSCRC RCC Materials
		• *Treating sickle cell disease: Is hydroxyurea right for your child?* English French Spanish
		• *Treating sickle cell disease: Is hydroxyurea right for you?* English French Spanish
		• Southeast
		  ○ Webinar: Hydroxyurea & Sickle Cell Children
		  ○ Webinar: Hydroxyurea & Sickle Cell Adults

Enhancement of Provider Awareness of Disease-Modifying Therapy Initiation and Dosing Materials	Providers, especially those who treat very few people living with SCD, such as primary care and advance practice providers, need resources to stay up-to-date on current SCD treatment guidelines. Pocket guides or guideline documents that outline initiation and dosing of HU are useful for physicians who may need a quick reference tool in the clinical setting. Continuing medical education (CME) webinars are another resource for primary care physicians who seek to increase their knowledge about HU.	Hydroxyurea Treatment for Adults (CME webinar) Hydroxyurea and Transfusion Therapy (ASH) **NE RCC materials** • Boston Medical Center Pediatric Hydroxyurea Dosing Guidelines • SiNERGe Webinar - Helping Patients Adhere to Hydroxyurea Therapy • SiNERGe WEBINAR: Patient Perspectives Part II **PSCRC Materials** Improving Preventive Care for Children With Sickle Cell Anemia: A Quality Improvement Initiative. (Cabana et al., 2020)

Immunizations

People living with SCD are at increased risk of invasive bacterial disease as well as more severe illness due to influenza, making vaccination an important preventive health measure. However, national data show that vaccination rates vary widely, both by age and vaccine type (National Academies of Sciences Engineering and Medicine, 2020). Three-quarters or more of patients with SCD nationally have received at least one of two recommended pneumococcal vaccines, while only 30-52 percent of patients have received both. Influenza vaccination ranges from 30-82 percent for pediatric SCD patients and 12-61 percent for adult SCD patients; only 17-24 percent of SCD patients received the meningococcal vaccine.

Tracking and documenting completion of immunization can be challenging. Patients may receive immunizations in multiple settings, yet ensuring up-to-date documentation in the patient's medical record is required to be able to administer needed immunizations. This can be especially challenging for SCD specialists who report that they are not always able to access primary care records or immunization registries — complete immunization records may not exist with any one provider. An additional challenge arises when patients receive immunizations outside the medical system, such as community locations (e.g., schools, local pharmacies).

The differing ways of getting immunizations means that valuable information may not be seamlessly transferred between systems, adding to fractured record-keeping. Establishing consistent, standard ways of tracking and sharing immunization data, such as reminder-recall systems, state-wide registries, and interoperable electronic health records (EHRs), are essential components of any effort to increase immunization coverage. However, technology alone is not sufficient. Optimized workflow and increased collaboration between primary care and specialty sites are also key components. Table 3 describes strategies and resources to support increasing the delivery of immunizations.

Table 3. Strategies and Resources to Increase Delivery of Immunizations

STRATEGY	RATIONALE	RESOURCES
Evidence-Based Guidelines and Protocols for Clinical Practice Support	The use of evidence-based guidelines is recommended. Guidelines should be developed using a systematic process, including literature review and grading of the evidence to address specific clinical questions developed by an expert panel (DeBaun, 2014; Yawn et al., 2014).	• NHLBI immunization guidelines • CDC Immunization Best Practices • Heartland/Southwest Vaccinations in SCD (see October 19, 2018)
Increased Use of Tracking Strategies	Increased use of tracking systems that can employ EHR reports and state-based immunization registries can improve completeness of immunization records, identify missing immunizations, and increase the number of fully immunized patients. Adjusting workflows can further ensure immunization completion.	One Heartland/Southwest site improved their immunization rates by creating a tracking template. Here is the pre-authorization template.

Transcranial Doppler

People who live with SCD are at increased risk for stroke, both silent stroke and overt stroke. Without prevention, approximately 10 percent of young adults by age 20 (Ohene-Frempong et al., 1998) and 24 percent of all patients by age 45 will have a stroke (Zétola, 2012). Adults who experience stroke have severe morbidity and high mortality rates. Transcranial Doppler (TCD) is a noninvasive ultrasound procedure that allows the clinician to clearly see how quickly blood is flowing through the brain during a period of time. High blood flow velocity is associated with an increased risk of stroke. This screening test is reliable, painless, and relatively inexpensive. Given these factors, TCD use with children aged 2-16 living with SCD is strongly recommended. The NHLBI reviewed two randomized-controlled trials and 50 observational studies in making their recommendations. The Stroke Prevention Trial in sickle cell anemia (STOP trial) demonstrated a 92% decrease in the rate of stroke in children with abnormal TCD when treated with monthly red blood cell transfusions compared to observation alone (Adams et al., 1998). While this is a potential life-saving test, uptake is low, with one study finding only approximately 45% of eligible children were screened (Raphael et al., 2008). Other studies have demonstrated variability in TCD rates, all with lower rates than desired, however. For example, one study showed 25 percent of children living with SCD aged 2-5 years received screening (Bundy et al., 2016) while another found 68 percent of 338 publicly insured children living with SCD (Eckrich et al., 2013) were screened. Overall, it appears that less than half of eligible children living with SCD receive appropriate TCD screening (National Academies of Sciences Engineering and Medicine, 2020).

WHAT IS TRANSCRANIAL DOPPLER (TCD)?

Transcranial Doppler (TCD) is a noninvasive ultrasound procedure that allows the clinician to clearly see how quickly blood is flowing through the brain during a period of time.

The Program has developed materials to guide clinicians on implementing systems for TCD completion as well as how to track completion.

Table 4. Strategies and Resources to Increase Delivery of Transcranial Doppler

STRATEGY	RATIONALE	RESOURCES
Evidence-Based Guidelines and Protocols for Clinical Practice Support	The use of evidence-based guidelines is recommended. Guidelines should be developed using a systematic process, including literature review and grading of the evidence to address specific clinical questions developed by an expert panel (DeBaun, 2014; Yawn et al., 2014).	• Risk of Stroke in Children with SCD (patient poster) • Southeast: Transcranial Doppler (TCD) Quality Improvement Protocol • Spreadsheet template to track TCD • Heartland/Southwest: TCD Guidelines (see January 18, 2019)

SICKLE CELL DISEASE TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Increasing Provider Knowledge Through Education

Model Protocol

Increasing Provider Knowledge Through Education

Children living with SCD are surviving into adulthood. The need for adult providers — primary care, advanced practice, and specialty providers — who are trained to effectively manage SCD continues to grow. While there are geographic areas where access to engaged, knowledgeable providers is available, there is still a shortage. Many people do not live within a reasonable commuting distance to comprehensive SCD care, and people who live in remote areas often have a difficult time reaching providers with significant expertise. Therefore, continued focus on expanding provider knowledge through educational opportunities is warranted.

EXPANDING PROVIDER KNOWLEDGE

Many people do not live within a reasonable commuting distance to comprehensive SCD care, and people who live in remote areas often have a difficult time reaching providers with significant expertise.

The RCCs employed several strategies to increase provider knowledge of SCD care. Traditional provider education methods were employed, including presentations at symposiums, grand rounds, national professional society meetings, and disseminating information through publications, and are recommended to continue. In addition, the table below reflects resources and protocols used by the RCCs. These provided opportunities to build engagement and awareness and enhance knowledge across broad audiences of providers. Focused training efforts, such as QI learning sessions and direct education training, offered opportunities for more in-depth learning about relevant content. Finally, RCCs employed telementoring for providers through the expansion of Project ECHO®. These were all important strategies to increase and spread provider knowledge about caring for people living with SCD.

RCCs expanded their use of Project ECHO ®. Sessions allow experts to mentor attendees, provide feedback on difficult patient cases, and share expertise. Project ECHO® presents opportunities for in-depth learning that can be applied directly to practice. This method of knowledge expansion is particularly important for rural and other populations who are underserved. All five Program RCCs initiated Project ECHO® replications as a method to share expertise and increase knowledge with the goal of ensuring all patients in their region could have access to high quality care. While Project ECHO® was not created for use with rare diseases such as SCD, it is a promising approach that will continue to be refined.

PROGRAM ECHO REPLICATION PROGRAMS

These are Program replications of Project ECHO®. Visit these websites to register, participate, and access curricula and session recordings.

- **Heartland/Southwest:** https://sicklecell.wustl.edu/scd-teleecho-clinic-164
- **Midwest:** https://sickleecho.org
- **Northeast:** https://www.hopkinsmedicine.org/Medicine/sickle/providers/index.html
- **Pacific:** https://pacificscd.org/project-echo/
- **Southeast:** In Development

More information about Project ECHO® in Appendix B

Table 5. Recommended Topics to Target for Improving Provider Knowledge Through Education

TOPICS	RESOURCES
General Information	• Educational Presentation on SCD for Primary Care Providers • *Health Maintenance and Management for Chronic Complications of Sickle Cell* • Effective Communication with Patients with Sickle Cell Disease • *Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action* (NASEM)
Screening for SCD	• *CDC Hemoglobinopathies: Current Practices for Screening, Confirmation, and Follow-up* (guidelines)
Pain Management	• Illinois SCDTDP Pain Chart • Wong-Baker FACES Pain Rating Scale© • Sickle Cell Treatment & Outcomes Research in the Midwest TeleECHO Clinics • Living with the Pain of SCD II (webinar) • Chronic Opioid Therapy & Sickle Cell Disease (webinar) • *Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014,* NHLBI (guidelines) • *Management of Acute Complications of Sickle Cell Disease* (pocket guide) • *Health Maintenance and Management of Chronic Complications of Sickle Cell Disease* (pocket guide) • Key Findings: CDC's Sickle Cell Data Collection Program: Data Useful in Describing Patterns of Emergency Department Visits by Californians with Sickle Cell Disease (article) • Intranasal Fentanyl: Fast Relief of Sickle Cell Pain (Massachusetts SCDNBSP, Boston Medical Center) • California SCDTDP ED IN Fentanyl • *Sickle Cell Pain in the Emergency Department: A Guide to Improving Care* • Emergency Room Pain Algorithm • SAFER card (patients to give to ER docs)

Medical Home/Care Coordination	• Individual Care Plans:
	  ○ Patient Needs Assessment
	  ○ SMART Phrase
	  ○ Care Coordination Screening Tool
	  ○ Care Coordination Checklist
	• Pre-appointment Planning Worksheet

Preventive Care	• *Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014,* NHLBI • Risk of Stroke in Children with SCD
Transitions from Pediatric to Adult Care	• *Making a Smooth Transition from Pediatric to Adult SCD Care: Eliminating Barriers, Enhancing Resources: Part I* (webinar) • *Making a Smooth Transition from Pediatric to Adult SCD Care: Eliminating Barriers, Enhancing Resources: Part II* (webinar) • *Recommended Curriculum for Transition from Pediatric to Adult Medical Care for Adolescents with Sickle Cell Disease: Suggested Topics, Methods, and Efficacy Measurements* (WISCH, PDF) • Please see the Compendium of Tools and Materials

SICKLE CELL DISEASE TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Increasing Use of Technology That Supports Healthcare Delivery

Model Protocol

Increasing Use of Technology That Supports Healthcare Delivery

Data Systems: Dashboards and Registries

Access to data is foundational to all high-performing QI efforts. Depending on available resources, data collection and reporting solutions can range from simple to complex and from relatively labor-intensive to highly automated. Also, levels of data-sharing can range from aggregate, summary data to more granular patient-level data. The extent to which both data collection and sharing can be done will be guided by institution governance and resource considerations.

This section will focus on the use of EHR systems for clinical care, as they are a powerful tool for improving care. The use of EHRs is now the norm, and electronic dashboards and registries are increasing in use each year. The data in these systems can and should be used to:

- Provide ongoing assessments of the quality of health and healthcare
- Identify patients with care gaps who need outreach
- Assist clinicians through decision-making support while seeing patients

EHR systems can be optimized to ensure efficient coordination of services, address care gaps, improve provider-patient communication, efficiently support medication refills, and remind clinicians about important actions. For example, incorporating transition-readiness assessments that are easily accessible at the appropriate time or programming smart phrases to sustain hydroxyurea counseling can positively impact completion rates (Cabana et al., 2020). But variation in site capacity to customize EHRs and differences between EHR products with regard to functionality can present challenges at many sites, especially if working collaboratively with other sites. EHR systems at sites may have slightly different formats that do not allow for easy export and upload into a centralized database. Also, data validation using a centralized cumulative repository is difficult without access to the source data. Nonetheless, as applicable, all teams that seek to improve care for people living with SCD will benefit from developing a strategy to optimize use of their EHRs.

All EHR systems support some degree of reporting for the underlying data in the system. Once developed, EHR reports can be run on a regular basis and be used to assess quality measures. Efforts to increase the sharing of reports for teams using the same EHR product could be expected to reduce the effort needed and broaden use of reporting.

The experience of teams during the Program reflect the experiences of others who are trying to use clinical data to support improvement work. Depending on local IT resources and access to data, some sites were limited to manual tracking of measures via desktop spreadsheets, while others were able to develop reports that could automate measurement and provide regular reports. Whatever the resources available, continuously gathering data to assess quality was seen as important throughout the Program, as were efforts to improve the degree to which electronic data could be accessed and used. The work that RCCs conducted to develop dashboards and registries reflected these sentiments.

Dashboards

Ongoing review of quality measures is an essential component of any QI activity. Quality dashboards that are updated regularly and easily accessible (embedded in the EHR system or shared externally) inform teams about their ongoing progress as they test and implement improvement strategies. For example, a pediatric site in the Pacific RCC used a healthcare quality dashboard embedded in the EHR system to monitor Program performance measures, including TCD completion and HU prescriptions, in a dynamic and interactive way. The dashboard allowed for real-time tracking of the effectiveness of interventions. Quality dashboards allow comparison between teams to identify best practices and areas in need of attention. Typically, a limited set of key quality measures are selected and updated regularly. Dashboard data can be supplied directly from the EHR system or via data extracted from IT departments. For teams where dashboards were available, they were viewed as an important communication platform that allowed them to assess whether the work they conducted was effective and to plan for new improvement activities.

Registries

Disease-specific registries can leverage data in multiple systems (EHR, patient surveys, manual entry) from various sites and have the potential to deliver optimized, curated data and reporting. Registries that focus on people with SCD have the potential to provide better access to data and ability to generate targeted quality measures, which can then be used to manage engagement though reminders and outreach. Insights learned from registry data can be used to provide education about patients and can help determine key metrics that align with specific regional programmatic efforts as well as the aims of the larger national initiative. When possible, reported quality measures for registries should be based on standard measure definitions and developed in a way that allows comparison between sites.

An important component for the data collections systems developed during the Program is one shared by most clinical systems: staff who are trained to manage the data system/applications and who understand the Program goals and quality measures. Ideal qualities for these staff include:

- Experience with gathering performance metrics in large, multisite environments (for multisite collaborations)
- Experience in end-user database design — collaborating with all principal investigators (PIs), data coordinators, and statisticians — incorporating site-specific elements in addition to common data elements (CDEs), and providing training manuals
- Ability to build databases, provide database tech support, refine the database, and provide data coordination/management services (one-stop shop)
- Ability to address missing data, validate data, and monitor performance for accurate reporting
- Ability to work with others at all levels, build relationships, be responsive, and provide excellent customer service

These tools, structures, and systems are critical to gaining access to timely and quality data:

- **Secure, Online Data Capture and Storage System**
  - Centralized database that is easily accessible and HIPAA-compliant
  - Inclusion of local PIs and site EHR representatives in sponsor discussions to see what data are possible to collect or if a program-specific query can be built, and to generate a cost estimate
  - Timely refinements based on user experiences and solicited feedback
  - Plan to back up data for verification and validation
- **Data Collection Strategy**
  - Agreement of sponsors and both regional and local PIs on CDEs and site-specific data collection and coordination strategies
  - Clarity around timelines and all data elements to be captured
  - Allowance of sufficient time for sites to submit data for cleaning
  - Validation of all data before final submission
- **Well-Specified Common Quality Measures and CDEs**
  - Team agreement on measure specifications before any programming begins. (Changes to databases cost time and money and the chance to capture some information may be lost.)
  - Central data dictionary linked to measure specifications for consistency across sites
- **Interface for Automated Data Sharing or Personnel to Enter Data**
  - Building program-specific queries or Application Programming Interfaces (APIs) to pull agreed upon EHR CDEs by specialized IT staff needs funding and should be part of pre-implementation activities
- **Analytic Functionality to Allow the Measurement and Sharing of Quality Measurement Results Over Time**
  - Analytic functionality of data in the registry

- **Legal Framework to Allow Sharing of Data Between Collaborating Institutions**
  - Data use and sharing agreements should be written into the protocol for use of a centralized program database.
- **Communications**
  - If working in a regional collaboration, planning scheduled time for meetings, initial training, updates, support, and regular communications builds a cohesive team approach to data collection. Lessons learned and issues can also be shared at these meetings.
  - Frequent, clear communication and emails to PIs and their coordinators about expected data submission timelines — with reminders — promote completion success.
  - Measures and Data Submission should be a standing agenda item on monthly meetings.
  - Regional and site performance metrics and outcome trends should be shared as updated with all partners throughout the length of a program. This promotes buy-in, points for discussion, and healthy competition.
- **Partnership**
  - Ideal local site initial planning team should include regional PIs, local PI, local data coordinator, regional data coordinator, local site EHR programmer/developer, and statistician (if applicable).
  - If payment for data is awarded, completion of data submissions must be linked and clearly articulated as a condition to sub-award payments by the lead institution.

Table 6. Resources for Dashboards and Registries

TOPICS	RESOURCES
Dashboards	• Pacific: Epic EHR Dashboard • Southeast: Sickle Cell Dashboard Infographic
Registries	https://covidsicklecell.org/ This is a COVID-19-specific SCD registry.

Table 7. Website Resources for SCD

TOPICS	RESOURCES
STORM (Sickle Cell Treatment and Outcomes Research in the Midwest)	http://sicklestorm.org/index.html
Pacific Sickle Cell Regional Collaborative (PSCRC)	http://pacificscd.org/
Heartland/Southwest Sickle Cell Disease Network	http://sicklecell.wustl.edu/
Sickle Cell Improvement in the Northeast Region through Education (SINERGe)	http://wepsicklecell.org/sinerge/
American Society of Hematology	https://www.hematology.org/research/sickle-cell-disease-and-sickle-cell-trait/sickle-cell-research-priorities
CDC	https://www.cdc.gov/ncbddd/sicklecell/index.html
NHLBI	https://www.nhlbi.nih.gov/health/educational/sickle-cell-awareness
NICHQ	http://sicklecell.nichq.org
Sickle Cell Disease Association of America	http://www.sicklecelldisease.org
American Sickle Cell Anemia Association	http://www.ascaa.org/
Sickle Cell Information Center at Emory	http://scinfo.org/
California Sickle Cell Resources	http://casicklecell.org/
Sickle Cell Disease Coalition	http://www.scdcoalition.org/
PCORI Sickle Cell Project	https://www.pcori.org/research-results/2019/disseminating-results-missed-scd-clinic-appointments-and-health-belief-model

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SICKLE CELL DISEASE
TREATMENT DEMONSTRATION
REGIONAL COLLABORATIVES PROGRAM

Compendium of Tools and Resources

NICHQ
National Institute for Children's Health Quality

SEPTEMBER 2021

Table of Contents

Provider and Clinician Education and Resources

Patients and Caregivers

Introduction

There are approximately 100,000 people in the United States living with sickle cell disease (SCD). Access to high quality healthcare and services with providers knowledgeable about SCD care is imperative. The Sickle Cell Disease Treatment Demonstration Regional Collaborative Program (SCDTDRCP) is mandated by Congress and funded through the U.S. Health Services Resources Administration (HRSA). More information about the Program can be found in Appendix B of the 2021 Congressional Report.

The materials listed in this compendium offer a range of tools, materials, and resources for providers and care teams in support of the strategies and activities they are using with patients and families to ensure the best quality of life for those with living with SCD. This Compendium of Tools and Materials for SCD resources is the companion piece to the SCDTDRCP 2021 Model Protocol, which details key strategies for improving patient access to high quality care and disease modifying treatments by knowledgeable providers.

A COMPENDIUM OF HELPFUL TOOLS

All the materials in this compendium are useful tools for providers treating SCD. Some are for sharing with patients to support awareness, self-care management, and shared decision-making. Others are protocols and best practices that primary care, emergent care, and specialists should know and use to serve their Program. During the SCDTDRCP 2018-2021 project, there were many resources and methods for spreading knowledge and awareness of SCD employed.

Wherever possible, the name of the provider or site contributing the resource is noted in parentheses after the title and link.

Provider and Clinician Education and Resources

Acute Care Resources

Education Materials and Tools for Providers and Clinics

- Acute Care Individual Times Tool (OH TDP)
- Staff training: Presentation on sickle cell disease to ED staff (OH TDP)
- Evaluation survey for presentation to ED staff (OH TDP)
- CRISIS: Experiences of People with Sickle Cell Disease Seeking Health Care for Pain Video (MD TDP)
- Pediatric ED: Pain Med Calculator (MA NBSP)
- Pediatric ED: Nursing In-service - 2012 (MA NBSP)
- Pediatric ED: Nursing In-service - 2014 (MA NBSP)
- APPT Scoring Guide (CA TDP)
- Fever and Acute Chest Syndrome Best Practices Principles Poster (CA TDP)
- Priapism in Sickle Cell Disease Best Practices Principles Poster (CA TDP)

NEED FOR ACUTE CARE RESOURCES

Due to limited general knowledge of sickle cell disease among physicians and lack of access to specialty care, many people with sickle cell disease access medical care through the emergency department. This group of resources and tools is most relevant to providers in the acute care or emergency department setting.

- CDC's Sickle Cell Data Collection Program: Data Useful in Describing Patterns of Emergency Department Visits by Californians with Sickle Cell Disease (SCD)
- ASH Pocket Guide: Management of Acute Complications of Sickle Cell Disease (University of Illinois, Midwest)
- Sickle Cell Disease Education: Care of Patients with Sickle Cell Disease for Primary Care Providers and Emergency Room Personnel (Indiana Hemophilia/Thrombosis Center, Inc. (Indiana State STORM)

Intranasal Fentanyl

- California SCDTDP ED Protocol for Intranasal Fentanyl
- ED Protocol Intranasal Fentanyl (CA TDP)
- Pediatric ED: Intransal Fentanyl Handout (MA NBSP)
- Kelly GS, Stewart RW, Strouse JJ, Anders JF. Intranasal fentanyl improves time to analgesic delivery in sickle cell pain crises. *American Journal of Emergency Medicine* 2018;36(7):1305-1307

Pain Action Plans

- Pain Action Plan - English (CA TDP)
- Pain Action Plan - Spanish (CA TDP)
- Adult ED: Individualized Pain Plan (MA NBSP)
- Individualized Pain Plan for Children with Sickle Cell Disease (PA NBSP/TDP)

Pain Assessment

- Sickle Cell Pain Chart (IL TDP)
- Pain Assessment Scale (NY NBSP)

Patient-Controlled Analgesia Pumps

- Pediatric ED: Patient Controlled Analgesia Handout (MA NBSP)

Patient Satisfaction Survey

- ED Patient Satisfaction Survey (MA NBSP)
- ED Patient Satisfaction Survey (CA TDP)

Standard Order Sets

- Acute Chest Syndrome Management Checklist (TN NBSP)
- Iron Overload Checklist (TN NBSP)
- Pain Checklist (created by TN NBSP)
- Stroke Checklist (created by TN NBSP)
- Pediatric ED: VOE Protocol (MA NBSP)
- Sickle Cell Pain Initial Order Set Moderate to Severe Pain (CA TDP)
- Fever in Sickle Cell Disease Algorithm (CA TDP)
- ED Algorithm for Sickle Cell Disease Pain Management (NJ TDP)
- Sickle Cell Disease Fever Pathway (Indiana State STORM)
- Pediatric Sickle Cell Pain Pathway (Indiana State STORM)

COVID-19

- Sickle Cell Disease School Letter with COVID updates (Children's Hospital MN, Midwest)
- COVID-19 anticoagulation recommendations in children (RCC, Pacific)
- Safety Measures Checklist for the Re-entry of School During COVID-19 (Norton Children's Medical Group Cancer Institute, Southeast)
- School Toolkit Cover Letter (Children's Hospital MN, Midwest)
- COVID-19 Sickle Cell Registry

Data Systems

- Epic EHR Dashboard Elements (Phoenix Children's Hospital – Phoenix, Pacific)
- Sickle Cell Trait EHR Counseling Prompt (MA NBSP)

Educational Materials and Tools for Providers and Clinics

- Adult Sickle Cell Curriculum (University of Arkansas for Medical Sciences, Heartland/Southwest)
- Evaluation and Management: Splenic Sequestration (Heartland/Southwest)
- EMBRACE Presentations (RCC, Southeast)
- Guidelines to Treat Acute Chest Syndrome (ACS) (Heartland/Southwest)
- Guidelines for Acute Management of Stroke (Heartland/Southwest)
- Guidelines for Aplastic Crisis (Heartland/Southwest)
- Guidelines to Treat Fever in SCD (Heartland/Southwest)
- Perioperative Guidelines for Patients with SCD (Heartland/Southwest)
- Guidelines for Emergency Treatment of Priapism (Heartland/Southwest)
- EMBRACE Presentation: Creating a New Sickle Cell Clinic: A Case Study: A few dedicated professionals discuss how and why they created a new county clinic in South Los Angeles
- Telemedicine Checklist (Medical College of Georgia – Augusta, Southeast)
- Module: Effective Communication with Patients with Sickle Cell Disease
- Enhancing Access to Care for Sickle Cell Disease in South Carolina (webinar)
- Poster Presentation: Provider Bias Training Poster (Jacobi Medical Center, Northeast)

Hydroxyurea

- American Society of Hematology's Hydroxyurea & Transfusion Therapy for Sickle Cell Disease Pocket Guide
- Boston Medical Center Pediatric Hydroxyurea Dosing Guidelines
- Hydroxyurea for Sickle Cell Disease: Indications, Dosing and Monitoring Guideline (STORM)
- SiNERGe Webinar: Helping Patients Adhere to Hydroxyurea Therapy
- SiNERGe Webinar: A Conversation about Hydroxyurea Part 2
- EMBRACE presentation: Laboratory and Morphologic Changes During Hydroxyurea Dose Escalation to MTD

WHAT IS HYDROXYUREA?

The drug hydroxyurea has been life-changing for those who are able to use it (it is not effective for all types of sickle cell disease), yet many people who are eligible are not using hydroxyurea. This group of resources have been used by providers to share information with patients about the benefits and risks of using hydroxyurea.

Hydroxyurea (continued)

- Journal Article: Improved Hydroxyurea Effect With the Use of Text Messaging in Children With Sickle Cell Anemia (TN TDP)
- Crosby, L. E., Shook, L. M., Ware, R. E. and Brinkman, W. B. (2015), Shared decision making for hydroxyurea treatment initiation in children with sickle cell anemia. *Pediatric Blood & Cancer, 62*: 184–185. doi: 10.1002/pbc.25124

Immunizations

- Vaccinations in SCD (see October 19, 2018)
- ACH Immunizations by Age
- NHLBI immunization guidelines (see pg. 29)

Journal Articles

- Shook LM, Farrell CB, Kalinyak KA, et al. Translating sickle cell guidelines into practice for primary care providers with Project ECHO®. *Med Educ Online*. 2016; 21:33616.
- Whiteman LN, Haywood C, Lanzkron S, Strouse JJ, Feldman L, Stewart RW. Primary Care Providers' Comfort Levels in Caring for Patients with Sickle Cell Disease. *South Med J*. 2015;108(9):531-536.

Pain Management

- ED Pain Management Protocol (Adult) (Jacobi Medical Center, Northeast)
- Pediatric Pain Medication Algorithm (Jacobi Medical Center, Northeast)
- Sickle Cell Pain in the Emergency Department: A Guide to Improving Care (NICHQ)

Point-of-Care Resources

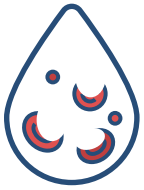
- Electronic evidence-based, primary care Platform: Dynamed Plus
- Online global medical news: Medscape
- Evidence-based, point-of-care clinical decision support system: Essential Evidence Plus
- STORM HU Clinical Decision Tool (contact storm@cchmc.org for information)

Project ECHO®

- Sickle Cell Treatment & Outcomes Research in the Midwest TeleECHO Clinics
- STORM TeleECHO Clinic Pamphlet
- Project ECHO for Sickle Cell Disease (SiNERGE)
- SCD Training and Mentoring Program (STAMP) (PSCRC)
- Washington University School of Medicine TeleECHO clinic
- Journal Articles
    - Shook LM, Farrell CB, Kalinyak KA, Nelson SC, Hardesty B, Saving K, Whitten-Shurney W, Panepinto J, Crosby LE, and Ware RE.(2016) Using Telementoring to Improve Sickle Cell Disease Care in the Midwest, *Medical Education Online*, Nov 24;21:33616. 27887664

Trait Resources

- Sickle Cell Trait Pre-Clinic Review (IL TDP)
- Sickle Cell Trait Provider CME Training (MA NBSP)
- Screening and Trait Counseling\Screening Algorithm (MA NBSP)



General Sickle Cell Disease

- Health Maintenance and Management of Chronic Complications of Sickle Cell (University of Illinois, Midwest)
- Management of Acute Complications of Sickle Cell Disease (University of Illinois, Midwest)
- Sickle Cell Issue Awareness Guide for Providers (University of Illinois, Midwest)
- SCDAA 2018-2019 Annual Report (Children's Hospital of Michigan – Michigan, Midwest)
- Educational Presentation on SCD for Primary Care Providers (Children's Hospital of Michigan – Michigan, Midwest)
- Setting Up a Comprehensive SDC Clinical Program (RCC, Southeast)
- Help Advance Care for Sickle Cell - Image (Heartland/Southwest)
- PhenX Toolkit
- REDCap
- Texas State Action Plan (Baylor College of Medicine / Texas Children's Hospital, Heartland/Southwest)
- Pacific RCC State Action Plans
- Heartland/Southwest State Action Plan
- Arkansas State Action Plan
- Iowa State Action Plan
- Kansas State Action Plan
- Louisiana State Action Plan
- Missouri State Action Plan
- Nebraska State Action Plan
- Oklahoma State Action Plan
- Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action (NASEM)
- Evidence Based Management of Sickle Cell Disease: Expert Panel Report, 2014 (NHLBI, NHI)

Regional Coordinating Centers SCDTDP 2014-2017

- Sickle Cell Treatment and Outcomes Research in the Midwest (STORM)
- Pacific Sickle Cell Regional Collaborative (PSCRC)
- Sickle Cell Improvement in the Northeast Region through Education (SiNERGe)

Websites

- American Society of Hematology (Sickle Cell)
- Centers for Disease Control and Prevention (Sickle Cell)
- National Heart Lung and Blood Institute
- National Institute for Children's Health Quality
- Sickle Cell Disease Association of America
- American Sickle Cell Anemia Association
- Sickle Cell Information Center at Emory
- Sickle Cell Disease Coalition
- California Sickle Cell Resources
- PCORI Sickle Cell Project
- Cincinnati Children's: Sickle Cell Trait
- Heartland Sickle Cell Disease Network

Health Maintenance and Tracking

- Adult Patient Tracking Log (IL TDP)
- Care Coordination Checklist (IL TDP)
- Form D.113B.1: Red Blood Cell Exchange and Depletion Standing Orders (RCC, Southeast)
- BEARS Sleep Screening Tool (Norton Children's Medical Group Cancer Institute, Southeast)
- Health Maintenance Form (NJ TDP)
- Pre-appointment Planning Worksheet (University of Illinois, Midwest)
- Sickle Cell Dashboard Infographics (RCC, Southeast)
- Sickle Cell Disease Clinic Worksheet (IL TDP)
- SMART Phrase: Quick summary of relevant sickle cell disease management facts (OH TDP)
- Colorado Learning Difficulties Questionnaire
- Transcranial Doppler (TCD) Quality Improvement Protocol (Medical College of Georgia – Augusta, Southeast)

Local Electronic Health Registries

- SCDTDP Minimum Data Set Data Dictionary (2014-2017)
- PSCRC Minimum Dataset
- PSCRC Enrollment and Annual Update Form
- PSCRC Abbreviated Provider Form
- COVID-19 Specific SCD Registry

Mental Health Screening

- Depression Checklist (TN NBSP)
- Mental Health Referral Protocol (MO TDP)
- Patient Referral Satisfaction Survey - Mental Health Services (MO TDP)
- Mental Health Referral Flowchart (MO TDP)
- Patient Health Questionnaire - Depression Screening (PHQ9 Copyright © Pfizer Inc. All rights reserved. Reproduced with permission. PRIME-MD® is a trademark of Pfizer Inc.)

Miscellaneous Journal Articles

- Ballas SK, Vichinsky EP. Is the Medical Home for Adult Patients with Sickle Cell Disease a Reality or an Illusion? *Hemoglobin*. 2015;39(2):130-133. doi:10.3109/03630269.2015.1023312.
- Hsu LL, Green NS, Donnell Ivy E, et al. Community health workers as support for sickle cell care. *Am J Prev Med*. 2016;51(1):S87-S98. doi:10.1016/j.amepre.2016.01.016.

Pain Management and Health Risks

- Inpatient Sickle Cell Pain Management Guidelines Presentation (Heartland/Southwest)
- Emergency Room Pain Algorithm (Heartland/Southwest)
- Yoga and Mindfulness for Pediatric Inpatients (Heartland/Southwest)
- Wong-Baker FACES Pain Rating Scale©
- Pain Treatment Guideline (Oregon Health and Science University, Northeast)

Patient Communication

- CAMC Sickle Cell Clinic Postcard (Charleston Area Medical Center / West Virginia, Northeast)
- AHA Sickle Cell - Save the Date 2019 (Phoenix Children's Hospital – Phoenix, Pacific)
- HRSA Telemedicine Patient Survey (Heartland/Southwest)
- WSCD 2020 Flyer (Phoenix Children's Hospital – Phoenix, Pacific)
- Pacific Sickle Cell Regional Collaborative Website (Pacific)
- Regional Updates Newsletters (Pacific)

Patient Education (Family)

- The Hope and Destiny Book
- Authorization for Release of Information Document (Children's Hospital MN, Midwest)
- Sickle Cell Trait Knowledge Tool (TN NBSP)
- The Talking Drums Project Community Survey (CA TDP)
- Readiness Assessment for Parents (Got Transition ®)
- Readiness Assessment for Youth (Got Transition ®)

Hydroxyurea

- Keeping You Healthy with Sickle Cell Disease. An educational kit addressing knowledge/beliefs on the use of hydroxyurea (MA NBSP)
- Treating sickle cell disease: Is hydroxyurea right for your child? (English) (PSCRC, STORM, Heartland SCD Network) (Spanish; French)

Quality Improvement

- Quality Improvement: Change Packet (RCC, Southeast)
- Cabana MD, Marsh A, Treadwell MJ, Stemmler P, Rowland M, Bender MA, Bhasin N, Chung JH, Hassell K, Rashid NN, Wong TE. Improving Preventive Care for Children With Sickle Cell Anemia: A Quality Improvement Initiative. Pediatric quality & safety. 2021 Jan;6(1).

Transition

- Transition Research Brief (Jacobi Medical Center, Northeast)
- Sickle Cell Transition Curriculum (RCC, Southeast)
- Got Transition® Toolkit (National Health Care Transition Family Advisory Group)
- Got Transition®
- Florida Health and Transition Services
- A Program of Transition to Adult Care for Sickle Cell Disease Journal Article (ASH)
- Recommended Curriculum for Transition from Pediatric to Adult Medical Care for Adolescents with Sickle Cell Disease, includes Topics, Methods, and Efficacy Measurements (PDF) Journal Article (WISCH)
- Journal Articles
    - Crosby, L. E., Quinn, C. T., & Kalinyak, K. A. (2015, April 1). A Biopsychosocial Model for the Management of Patients With Sickle-Cell Disease Transitioning to Adult Medical Care. *Advances in Therapy*. Springer Healthcare.
    - Treadwell M, Johnson S, Sisler I, et al. Development of a sickle cell disease readiness for transition assessment. *Int J Adolesc Med Health*. 2016;28(2):193-201. doi:10.1515/ijamh-2015-0010
    - Treadwell M, Johnson S, Sisler I, et al. Self-efficacy and readiness for transition from pediatric to adult care in sickle cell disease. *Int J Adolesc Med Health*. 2015;2015(4):381-388. doi:10.1515/ijamh-2015-0014.

Webinar Series

- Making a Smooth Transition from Pediatric to Adult SCD Care: Eliminating Barriers, Enhancing Resources Part I & II (Webinar Series) (SiNERGe)
- CDC's Sickle Cell Data Collection Program: Data Useful in Describing Patterns of Emergency Department Visits by Californians with Sickle Cell Disease (SCD) (PSCRC)
- Genotyping in Hemoglobin Disorders Webinar (STORM)

Patients and Caregivers

EDUCATION AND RESOURCES

People living with sickle cell disease often have multiple and complex medical needs, so it is ideal to be able to coordinate care among various specialties. This group of resources is useful for providers, patients, caregivers and representatives of community-based organizations who are involved in coordinating care for patients.

COVID-19

- COVID-19 Handout (Heartland/Southwest)
- COVID-19 Infographic (RCC, Southeast)
- School Reopening MARAC Recommendations (Children's Hospital MN, Midwest)
- COVID Tip Sheet (Children's Hospital MN, Midwest)
- School is Closed Tip Sheet (Children's Hospital MN, Midwest)

Health Maintenance and Tracking

- Care Coordination Screening Tool (IL TDP)
- Well Sickle Checklist (NY NBSP)
- Patient Event Diary (NY NBSP)
- Patient Needs Assessment (IL TDP)
- Sickle Cell Daily Activity Worksheet (Heartland/Southwest)
- Sickle Cell Disease Youth Acute Pain Functional Ability Questionnaire (Heartland/Southwest)

Living with SCD (Patients)

- Yoga and Mindfulness (Heartland/Southwest)
- Patient Empowerment Toolkit and Instructions for Use
  For questions, contact Patient Support(Children's Hospital of Michigan – Michigan, Midwest)
- Tips for Supporting Students with Sickle Cell Disease (CDC)
- Living Well with Sickle Cell Disease: Tips for Healthy Living (English; Spanish) (CDC)
- 5 Tips to Prevent Infection (CDC)
- SAFER Card (Children's Hospital of Michigan – Michigan, Southwest)
- What You Should Know About Sickle Cell Disease: Nine Steps to Living Well with Sickle Cell Disease in College (CDC)

Miscellaneous Journal Articles

- Ballas SK, Vichinsky EP. Is the Medical Home for Adult Patients with Sickle Cell Disease a Reality or an Illusion? *Hemoglobin*. 2015;39(2):130-133. doi:10.3109/03630269.2015.1023312.
- Hsu LL, Green NS, Donnell Ivy E, et al. Community health workers as support for sickle cell care. *Am J Prev Med*. 2016;51(1):S87-S98. doi:10.1016/j.amepre.2016.01.016.

Patient Communication

- Local Resources for Patients with SCD (Phoenix Children's Hospital – Phoenix, Pacific)
- Pacific Sickle Cell Regional Collaborative Website (Pacific)
- Regional Updates Newsletters (Pacific)

Patient Education

- Pregnancy During Sickle Cell (Heartland/Southwest)
- Risk of Stroke in Children with SCD (Southeast)
- Sickle Stroke Screen Pamphlet (Southeast)
- Disease Education Materials (Norton Children's Medical Group Cancer Institute, Southeast)
- Sickle Cell Disease: Avascular Necrosis (AVN) (Children's Hospital St. Louis, Heartland SCD Network)
- Sickle Cell Disease: Bedwetting (nocturnal enuresis) (Children's Hospital St. Louis, Heartland SCD Network)
- Sickle Cell Disease: Retinopathy (Children's Hospital St. Louis, Heartland SCD Network)
- Sickle Cell Disease: Gallstones (Children's Hospital St. Louis, Heartland SCD Network)
- Sickle Cell Disease: Priapism (Children's Hospital St. Louis, Heartland SCD Network)
- SCD Fact Sheet: What you should know about sickle cell disease (English) (CDC) (French)

Hydroxyurea

- Hydroxyurea Brochure (Children's Hospital of Michigan – Michigan, Midwest)
- Hydroxyurea White board (Children's Hospital of Michigan – Michigan, Midwest)
- St. Jude Informative Hydroxyurea Pamphlet (St. Jude, Southeast)
- Hydroxyurea Recurring Brochure (Cincinnati Children's Hospital, Midwest)

Screening and Trait Resources

Educational and Counseling Strategies

- Genes for Teens Brochure (TN TDP)
- Genes for Parents of Children with Sickle Cell Disease (TN TDP)
- Trait Counseling Educational Booklet and Presentation (MO TDP)
- Sickle Cell Trait Counseling Handout (MA NBSP)
- Sickle Cell Trait Presentation for the Community (MA NBSP)
- A Parents' Guide to Sickle Cell Disease (MA NBSP)
- A Parents' Guide to Sickle Cell Trait (MA NBSP)
- Get Screened to Know Your Sickle Cell Status (English) (CDC) (Spanish; French)
- What you should know about Sickle Cell Trait (English) (CDC) (Spanish; French)
- What you should know about SCD and Pregnancy (English) (CDC) (Spanish; French)

SICKLE CELL TRAIT

Though carriers of the sickle cell trait do not have the same symptoms as those with sickle cell disease, it is important for individuals to know their status and how it can impact them and their families. Individuals often become aware of their status through community health fairs or during pregnancy, when screening for sickle cell disease and general education are offered. This group of resources can be used by providers to educate patients about sickle cell disease, sickle cell trait, and screening.

- Cincinnati Children's Hospital: Sickle Cell Trait (STORM)
- Sickle Cell Disease: Contraception (Children's Hospital St. Louis, Heartland SCD Network)
- Sickle Cell Disease: Pregnancy and Sickle Cell Disease (Children's Hospital St. Louis, Heartland SCD Network)

Pre- and Post-Tests

- Pre- and Post-Test for Genetic Counseling and Education (IL NBSP)

Other

- Sickle Cell Trait Toolkit (CDC)

Transition

- Health Care Transition Timeline for Parents and Caregivers (Got Transition ®)
- St. Jude E-learning Transition Platform (Southeast)
- Pediatric to Adult Health Care Transition: A Family Toolkit (Got Transition ®)
- Sickle Cell Transition from age 13 to 15 (Johns Hopkins All Children's Hospital, Northeast)
- Sickle Cell Transition from age 16 to 18 (Johns Hopkins All Children's Hospital, Northeast)
- Sickle Cell Transition from age 19 to 21 (Johns Hopkins All Children's Hospital, Northeast)
- Transition Binder (Children's Hospital MN, Midwest)

Webinar Series

- Webinar: Blood Transfusions & Iron Overload (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Understanding Your CBC (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Blood Transfusions & Blood Matching (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Blood Transfusions: How, When, Why (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Hydroxyurea & Sickle Cell Adults (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Hydroxyurea & Sickle Cell Children (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Diagnosis & Treatment of Acute Chest Syndrome (Sickle Cell Community Consortium (SC3), Southeast)
- Webinar: Bone Health & Avascular Necrosis (Sickle Cell Community Consortium (SC3), Southeast)

These resources were developed through the Sickle Cell Treatment Demonstration Regional Collaboratives Program (2017-2021). As the National Coordinating Center for this regional collaborative, NICHQ freely shares guidance, tools, and resources that teams from around the country have created, tested, or used to improve care for patients with sickle cell disease.

Source: National Institute for Children's Health Quality (NICHQ)

Published: 2021