OKLAHOMA
Health Care Authority

Independent Evaluation of Sickle Cell Disease Management within the SoonerCare Population

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LEGISLATIVE REPORT IN COMPLIANCE WITH SB 1467

Prepared by the Pacific Health Policy Group for:

State of Oklahoma
Oklahoma Health Care Authority

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INDEPENDENT EVALUATION

This independent evaluation of the SoonerCare program's performance in covering members with Sickle Cell Disease was conducted by The Pacific Health Policy Group (PHPG). PHPG is solely responsible for the analysis and findings presented in this report.

PHPG is a national consulting firm with locations in the states of Arizona, California, Illinois, Oklahoma and Vermont. PHPG specializes in the development and evaluation of programs to serve Medicaid beneficiaries with special health care needs.

PHPG wishes to acknowledge the cooperation of the Oklahoma Health Care Authority in obtaining the necessary data for completion of the evaluation. PHPG also wishes to acknowledge the cooperation of Supporters of Families with Sickle Cell Disease and the State's two centers-of-excellence (Jimmy Everest Center at Oklahoma University and Saint Francis Hospital).

TABLE OF CONTENTS

COMMONLY-USED ABBREVIATIONS & ACRONYMS

ABD	Aged, Blind, Disabled
CCU	Chronic Care Unit
DUR	Drug Utilization Review
EPSDT	Early and Periodic Screening, Diagnosis and Treatment
FDA	Food and Drug Administration
FPL	Federal Poverty Level
HAN	Health Access Network
HbSC	Hemoglobin C
HbSS	Hemoglobin S
HIE	Health Information Exchange
HMP	Health Management Program
HRSA	Health Resources and Services Administration
MCE	Managed Care Entity
OHCA	Oklahoma Health Care Authority
OSDH	Oklahoma State Department of Health
OSU	Oklahoma State University
OU	Oklahoma, University of
PCCM	Primary Care Case Management
PCMH	Patient Centered Medical Home
PCP	Primary Care Provider
PHE	Public Health Emergency
RFP	Request for Proposals
SCD	Sickle Cell Disease
SCT	Sickle Cell Trait
SDOH	Social Determinants of Health
SFY	State Fiscal Year

A. EXECUTIVE SUMMARY

Introduction

Sickle Cell Disease (SCD) is the most prevalent inherited blood disorder in the United States. There are an estimated two million Americans with the sickle cell trait (SCT), meaning that the individual inherited the sickle cell gene from one parent. There are approximately 100,000 Americans who have inherited the SCD gene from both parents and have been diagnosed with sickle cell anemia or another disease within the SCD group.

Sickle Cell Disease is present at birth, with symptoms often appearing in the first year of life and worsening over time. Children and adults with SCD are at greater risk of infection than the general population, including a heightened risk of pneumonia. Children and adults with SCD also can be at heightened risk for stroke, among other complications.

Many persons with SCD receive health care services through their state Medicaid program. In Oklahoma, the SoonerCare Program, in a typical year, covers between 400 and 500 members with SCD.

SoonerCare members with SCD are not evenly distributed throughout the State. Most reside in Oklahoma and Tulsa Counties, each of which is home to over 100 persons. The next most populated counties are Cleveland, Comanche and Muskogee, each with between 10 and 30 persons. There are 31 counties with at least one, but fewer than 10 members with SCD; 41 counties have no members with SCD.

Most members with SCD are enrolled in SoonerCare Choice, the OHCA's primary care case management model. In April 2024, the majority of SoonerCare Choice members will be enrolled into Managed Care Entities under the OHCA's new SoonerSelect model.

A variety of new prescription drugs and interventions have been developed for SCD treatment in recent years, raising life expectancy for those with some form of the condition. Despite these advances, SCD can be a devastating and difficult-to-manage condition for the patient and his or her family. In addition to other health risks and complications, persons with SCD may experience severe pain crises brought on by clotting of the abnormally shaped red blood cells.

Patients in crisis often require intensive and continuous opioid-based pain medications that must be administered parenterally (e.g., by intravenous method). The medications must be provided either in an emergency room or inpatient setting, where the patient can be monitored and the dosage increased as necessary to achieve pain relief. One SoonerCare member with SCD recently described the experience of an acute pain crisis as being, *"like shards of glass running through your system."*

SB 1467 Study Scope

During the 2022 regular session, the Oklahoma Legislature enacted Senate Bill (SB) 1467, which was signed into law by the Governor on May 2, 2022. Section 1A of SB 1467 directed the OHCA to:

> *"… conduct an annual review of all medications and forms of treatment for sickle cell disease and services for enrollees with a diagnosis of sickle cell disease. The purpose of the annual review is to determine if the available covered medications, treatments, and services are adequate to meet the needs of enrollees with a diagnosis of sickle cell disease, and whether the Authority should seek to add or recommend additional medications, treatments, or services."*

The OHCA retained the Pacific Health Policy Group (PHPG) to conduct an independent evaluation in accordance with SB 1467 requirements. PHPG is a national consulting firm that specializes in development and evaluation of programs to serve Medicaid populations with special needs.

PHPG organized the study scope around the Legislature's specific areas of inquiry. The evaluation methods similarly were selected to obtain the data necessary to inform findings and recommendations across the areas defined in SB 1467. They included: interviews with members (through a structured survey), providers and program stakeholders; analysis of Medicaid eligibility and paid claims data; and review of national best practices, among other activities.

PHPG applied the data and related analysis toward answering the following questions:

- *Do SoonerCare members with SCD have access to all necessary services, including access to knowledgeable Patient Centered Medical Home (PCMH) providers?*
- *Do Oklahoma emergency room providers have the appropriate training and resources to care for members in crisis?*
- *Do SoonerCare members with SCD have appropriate supports to navigate the health care system?*
- *How can the program be strengthened?*

Do Members Have Access to All Necessary Services?

Findings

Individuals with SCD often require support from multiple specialties, with Hematology typically serving as the nexus for their care. The SoonerCare program is open to all licensed and qualified physicians in the State.

Oklahoma has two recognized "centers of excellence" for treatment of persons with SCD, both of which serve SoonerCare members. They are the Jimmy Everest Center at Oklahoma University (OU) Children's Hospital in Oklahoma City and the pediatric hematology program at Saint Francis Health System in Tulsa. The OU program was created in 1993 and has served approximately 200 pediatric patients over the past two years. The Saint Francis program serves about 90 patients at any point in time.

SoonerCare members with SCD also have access to non-clinical assistance through Supporters of Families with Sickle Cell Disease, a comprehensive community-based organization serving individuals and families living with sickle cell and thalassemia disease and trait in Oklahoma. The organization is based in Tulsa but works on behalf of families throughout the State.

SoonerCare providers generally are satisfied with scope-of-coverage, from an insurance perspective. This is unsurprising, given that Medicaid coverage generally is comparable to commercial insurance in scope. However, providers at the centers-of-excellence stated that transportation can be a burden, particularly for members traveling long distances.

Specialist providers also questioned the rule under SoonerCare Choice that all referrals must originate with the member's PCMH. The centers-of-excellence, for example, must route patients back to their PCMH when a referral to an outside specialty is needed, potentially delaying care. The additional step keeps the PCMH informed but there are information sharing options short of a patient visit that would accomplish the same ends.

SoonerCare members with SCD are satisfied with some aspects of care but are careful to distinguish between specialists and providers at the centers-of-excellence and the broader PCMH community. In the words of one survey respondent, "*(My son's) Hematologist and Oncologist are good. His regular doctor does not know anything about SCD though."*

Recommendations

1. The OHCA, in conjunction with SoonerSelect MCEs, centers-of-excellence and Supporters of Families with SCD, should conduct a coordinated educational campaign targeting PCMH providers in counties with SCD members. PCMH providers could be offered continuing education credits for participating.

2. The OHCA and SoonerSelect MCEs should consider having a process for allowing qualified specialists to make referrals, while keeping PCMH providers informed, either directly or through the State's Health Information Exchange (HIE).

Do ER Providers Have Appropriate Training and Resources?

Findings

SoonerCare members with SCD who experience a pain crisis must be treated in a hospital setting, either in the emergency room or as an inpatient. Medications are administered parenterally and require continuous monitoring.

In State Fiscal Year (SFY) 2022, 327 out of 487 members with SCD (67.1 percent) had at least one emergency room visit. On average, members with SCD had approximately four emergency room visits per year.

The emergency rooms at OU Health Sciences Center and Saint Francis have evidence-based protocols for treatment of patients in crisis and providers are familiar with how to treat the condition. However, the majority of emergency room physicians may see only one or two cases a year. Many surveyed members and Town Hall participants expressed frustration with their emergency room experiences.

One survey respondent said, *"I honestly do not think the doctors in the emergency room have any idea of the excruciating pain – pain the rest of us will never know – these kids are going through. Yet, the doctors act like we are in there seeking drugs for no reason."*

Another said, "*"I cannot get treatment for my Sickle Cell disease…I stopped even going to the E.R. because it is a waste of time. Sometimes I just curl up in my bed and cry from the pain."*

A best practice described in literature and recommended by Supporters of Families with Sickle Cell Disease is creation of a pain management action plan. The plan is a written description from the patient's Hematologist that outlines his or her condition, needs and recommended course-of-care when in crisis. Patients with an action plan have a better chance of receiving timely care when they arrive in an unfamiliar emergency room. The action plans also could be uploaded to the State HIE for ready access from any hospital.

Recommendations

1. The OHCA, in collaboration with centers-of-excellence and Supporters of Families with Sickle Cell Disease, should undertake an educational campaign to increase knowledge of evidence-based protocols for treatment, including those available

through the American Society of Hematology and Centers for Disease Control and Prevention.

2. The OHCA, SoonerSelect MCEs and advocacy community should collaborate on outreach to members with SCD and Hematology community to facilitate creation of plans. Emergency room providers also can be educated on their efficacy and importance.

Do Members Have Appropriate Supports to Navigate the System?

Findings

Individuals with complex/chronic disease such as SCD often require care from multiple medical specialties, as well as behavioral health services to cope with what is a life-long condition. Navigating the health care system without support can lead to fragmented care or gaps in care, as well as patient discouragement.

Medicaid beneficiaries often face additional, non-clinical hurdles to accessing care. These factors, known as "social determinants of health" (SDOH) can include housing insecurity, food insecurity, difficulty making utility payments and lack of reliable transportation, among others. A person with significant SDOH needs may, by necessity, regard his or her health care, particularly preventive services, as a lesser priority.

One recognized best practice for managing complex care needs is through establishment of a member-centered interdisciplinary care team. The team typically includes representatives from all specialties relevant to the individual's health needs, as well as a designated care manager (nurse or social worker) to coordinate the team's activities. As suggested by its name, the team places the member at its center, and she or he retains autonomy for choosing the preferred course of care.

Individuals enrolled with an interdisciplinary care team typically receive a comprehensive assessment, followed by creation of a care plan that addresses both clinical and non-clinical (SDOH) priorities. SDOH needs may be managed by a Community Health Worker trained for this task.

The interdisciplinary care team model also is well-suited for facilitating a member's transition from pediatric to adult care. The team can assist the member in making the transition and can itself evolve, in terms of composition, from pediatric to adult care providers.

Another best practice is use of mobile app technology as a means of monitoring a member's health status and adherence to preventive care guidelines. Recent research

indicates the technology can be effective in reducing acute care utilization among persons with SCD.

There are multiple pathways through which SoonerCare members with SCD can receive care management today. Members who are seen at Jimmy Everest have access to an interdisciplinary care team in accordance with best practices. In addition to addressing current needs, the team assists members to prepare for the transition from pediatric to adult care and coverage. The OHCA administers several programs through which agency nurses or vendor staff provide care management to members with complex/chronic conditions, including a portion of the population with SCD.

SoonerSelect will expand access to interdisciplinary care management. MCEs will be required to offer an initial health screening to all new enrollees, and to perform a comprehensive clinical and SDOH assessment on those identified as having special needs, a category that would include members with SCD. The assessment will be used to develop a comprehensive, interdisciplinary care plan, to be overseen by a designated care manager.

Recommendations

1. The OHCA should ensure that current and future care management systems emphasize the importance to members of having a comprehensive care/action plan that addresses the member's complete care needs, both current and future for members transitioning from pediatric to adult coverage.

2. The OHCA should strive to make available interdisciplinary care management, where appropriate, by coordinating with the Jimmy Everest Center, SoonerCare HMP and SoonerCare HANs on behalf of ABD beneficiaries. (All SoonerSelect enrollees will have access to interdisciplinary care teams.)

3. The interdisciplinary care teams should be tasked with facilitating the transition of adolescents from pediatric to adult coverage and care.

4. The OHCA should explore use of a mobile app for members with SCD, either directly or through its contractors.

Conclusion

All study recommendations are actionable in the next twelve to eighteen months. The second annual report, due to the Legislature in January 2024, will provide an update on the status of the recommendations.

B. GENERAL BACKGROUND INFORMATION

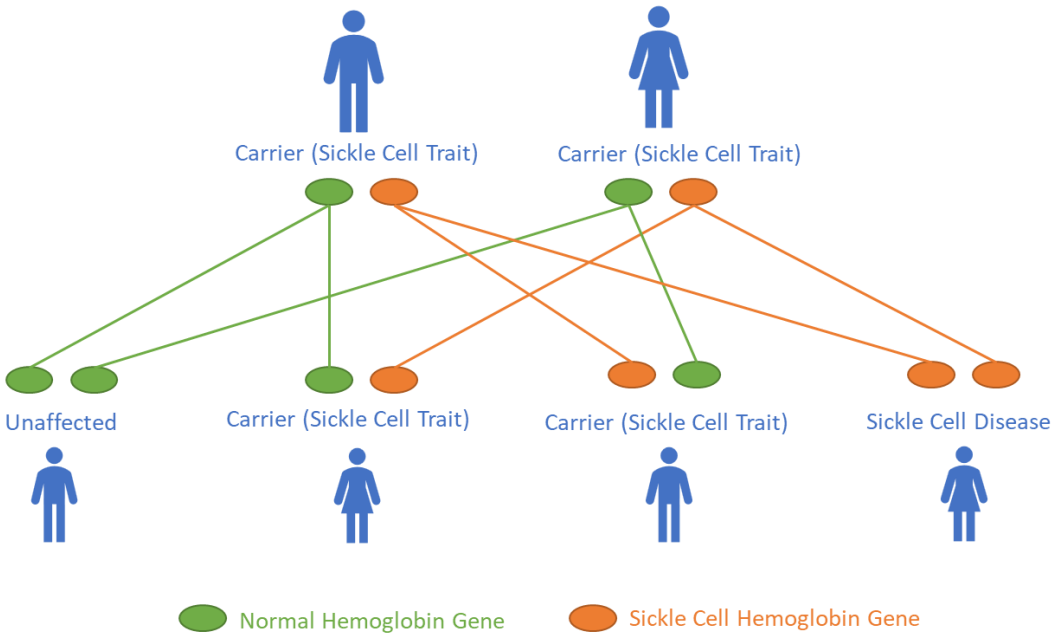
1. *Introduction*

Sickle Cell Disease Types and Prevalence

Sickle cell disease (SCD) refers to a group of blood disorders, usually inherited, of which Hemoglobin S (HbSS), also known as sickle cell anemia is the most common. SCD is concentrated within the African American community, where it occurs in one of every 365 births[1], making it the most prevalent inherited blood disorder in the United States.

There are an estimated two million Americans with the sickle cell trait (SCT) in the United States, meaning that the individual inherited the sickle cell gene from one parent. SCT occurs in approximately one of every 13 African American births. A child whose parents carry the SCD gene has a three-in-four chance of being born either with SCT or SCD (Exhibit B-1).

Exhibit B-1 – Sickle Cell Trait and Disease Risk

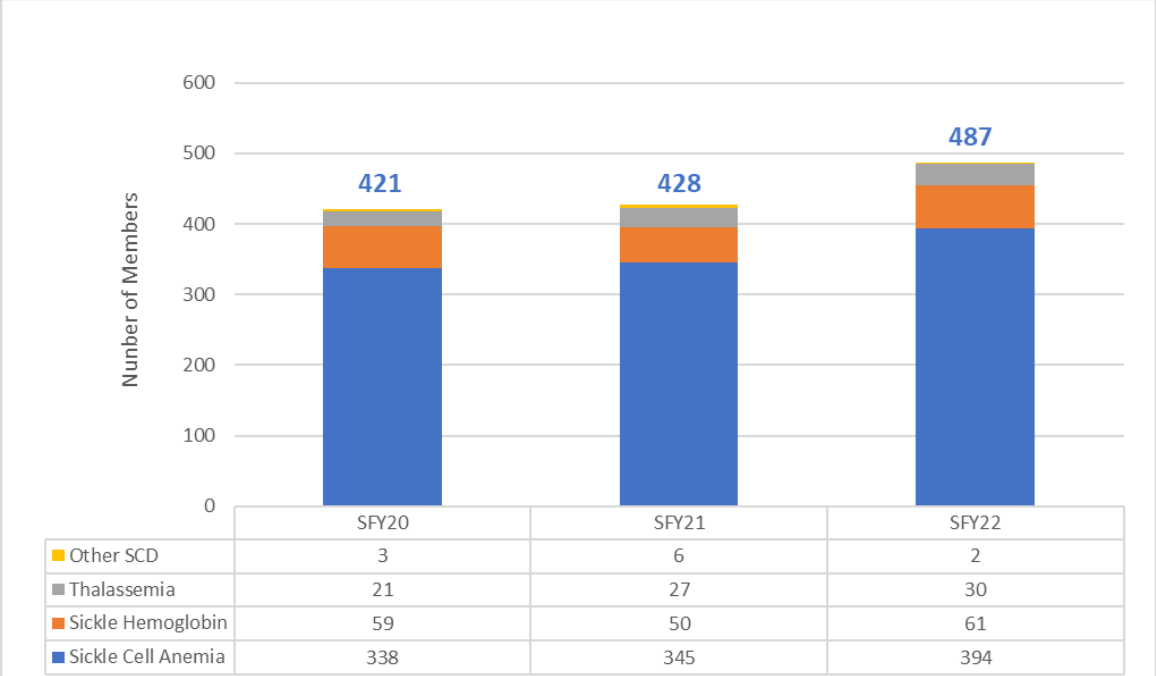


[1] Source: Data & Statistics on Sickle Cell Disease | CDC. SCD occurs in one of every 16,300 births to Hispanic Americans and less frequently among individuals of Asian, Mediterranean and Middle Eastern lineage.

There are approximately 100,000 Americans who have inherited the SCD gene from both parents and have been diagnosed with sickle cell anemia or another disease within the SCD group[2]. Other SCD conditions include Hemoglobin C (HbSC) and HbS beta thalassemia, as well as several rarer types. Sickle cell anemia, in which an abnormal form of hemoglobin causes red blood cells to become rigid and sickle-shaped, is usually the most severe form of SCD.

Many persons with SCD receive health care services through their state Medicaid program. During each of the three most recent state fiscal years (SFY 2020 – SFY 2022[3]), the SoonerCare program covered between 400 and 500 members with SCD[4] (Exhibit B-2).

Exhibit B-2 – SoonerCare Members with SCD by State Fiscal Year



	SFY20	SFY21	SFY22
Other SCD	3	6	2
Thalassemia	21	27	30
Sickle Hemoglobin	59	50	61
Sickle Cell Anemia	338	345	394

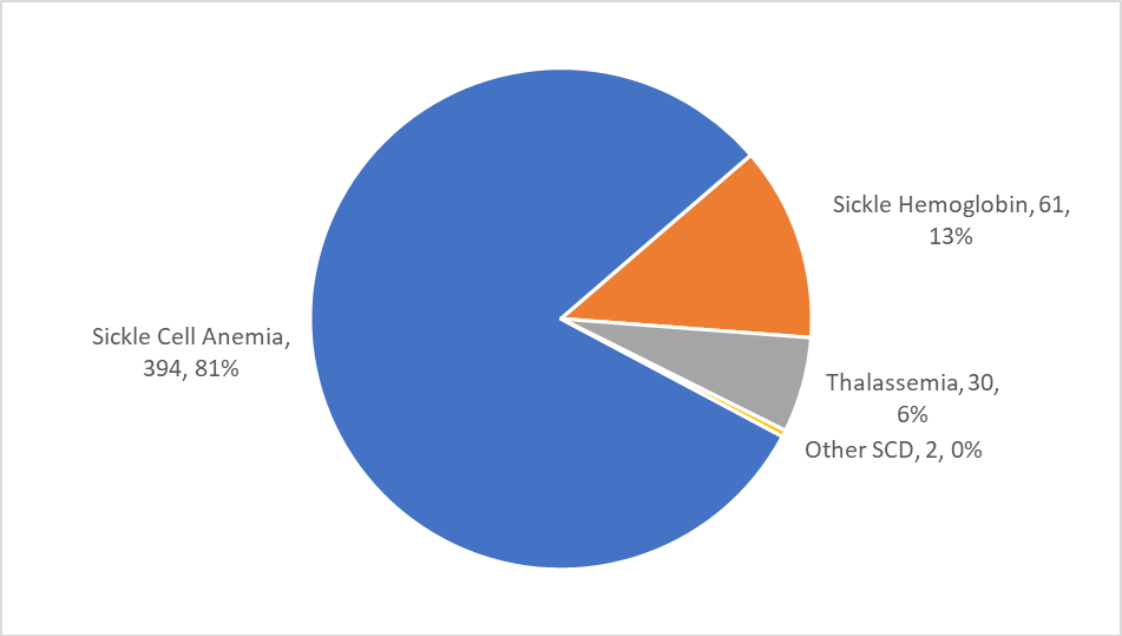
Over 80 percent of SoonerCare members with SCD were diagnosed with sickle cell anemia while other SCD conditions occurred with less frequency (Exhibit B-3 on the following page).

[2] Sickle cell disease: MedlinePlus Genetics

[3] State fiscal years run from July to June.

[4] Precise counts on the total number of Oklahomans with SCT and SCD are not readily available. Supporters of Families with Sickle Cell Disease, a leading community-based organization in the State, estimates there are 40,000 Oklahomans with the sickle cell trait and 1,500 families with one or more members who has been diagnosed with sickle cell anemia or another SCD. See: Sickle Cell Oklahoma – Supporters of Families with Sickle Cell Disease

Exhibit B-3 – SoonerCare Members with SCD by Category (SFY 2022)



Sickle Cell Disease Characteristics and Treatments

Sickle Cell Disease is present at birth, with symptoms often appearing in the first year of life and worsening over time. Children and adults with SCD are at greater risk of infection than the general population, including a heightened risk of pneumonia. In addition to being vaccinated, children with sickle cell anemia and severe forms of thalassemia are recommended to take penicillin daily until at least age five.

Children and adults with SCD also can be at heightened risk for stroke, which is identifiable through a special type of ultrasound (transcranial Doppler ultrasound). Stroke risk can be reduced through administration of frequent blood transfusions. Transfusions also are used to address episodes of severe anemia.

The transfusions themselves can cause side effects such as iron overload, which pose the risk of damage to the heart, liver, kidneys and other organs. Blood transfusions typically are accompanied by iron chelation therapy to reduce excess iron in the body[5].

A variety of prescription drugs have been developed for SCD treatment and can be used for young children and adolescents. These include, among others:

Hydroxyurea – this medication was approved in the 1980s and reduces the development of abnormally-shaped red blood cells. It can be prescribed starting at age two.

[5] This is not an exhaustive listing of SCD-related complications, which can be damaging to many body systems.

L-glutamine – this is an amino acid that supports the body's fight against infections. It also helps to reduce damage to blood cells. L-glutamine treatments can be prescribed starting at age five.

Voxelotor – this medication helps to restore red blood cells to their normal shape and can be prescribed starting at age four.

Crizanlizumab – this medication reduces the risk of blood cell clumping or clotting. It can be prescribed starting at age 16.

There also are new medications in clinical trials or awaiting FDA approval. One recently-developed medication – Adakveo – can reduce the frequency of pain crises in older adolescents and adults; another – Oxbryta – can be used to lower the risk of anemia in adolescents and adults. Medicaid covers all FDA-approved treatments, although some medications require prior approval on a case-by-case basis.

Persons with SCD also may be candidates for bone marrow or stem cell transplants. These procedures offer the potential for a cure but also have high risks and potential serious side effects. They also require a donor who is a close genetic match to the patient, such as a sibling.

The introduction of new medications and treatments in recent decades has resulted in reduced mortality rates among younger persons with SCD. Nationally, from 1979 to 2017, the median age at death increased from 28 years to 43 years. Over that same time period, SCD-related death rates among Black children younger than five years of age declined, from 2.05 deaths per 100,000 to 0.47 deaths per 100,000.

Despite these advances, SCD can be a devastating and difficult-to-manage condition for the patient and his or her family. In addition to other health risks and complications, persons with SCD may experience severe pain crises brought on by clotting of the abnormally shaped red blood cells.

Patients in crisis often require intensive and continuous opioid-based pain medications that must be administered parenterally (e.g., by intravenous method). The medications must be provided either in an emergency room or inpatient setting, where the patient can be monitored and the dosage increased as necessary to achieve pain relief. One SoonerCare member with SCD recently described the experience of an acute pain crisis as being, "*like shards of glass running through your system*."[6]

[6] Member participated in a November 30 Town Hall organized by Supporters of Families with Sickle Cell Disease. Chapter two includes more information on the meeting and quotes from participants.

Sickle Cell Disease – Barriers to Care

Patients and families with SCD face numerous potential health disparities/barriers to care. Patients living outside of major metropolitan areas may not have local access to a Hematologist with specialized knowledge of the condition, necessitating lengthy travel for care.

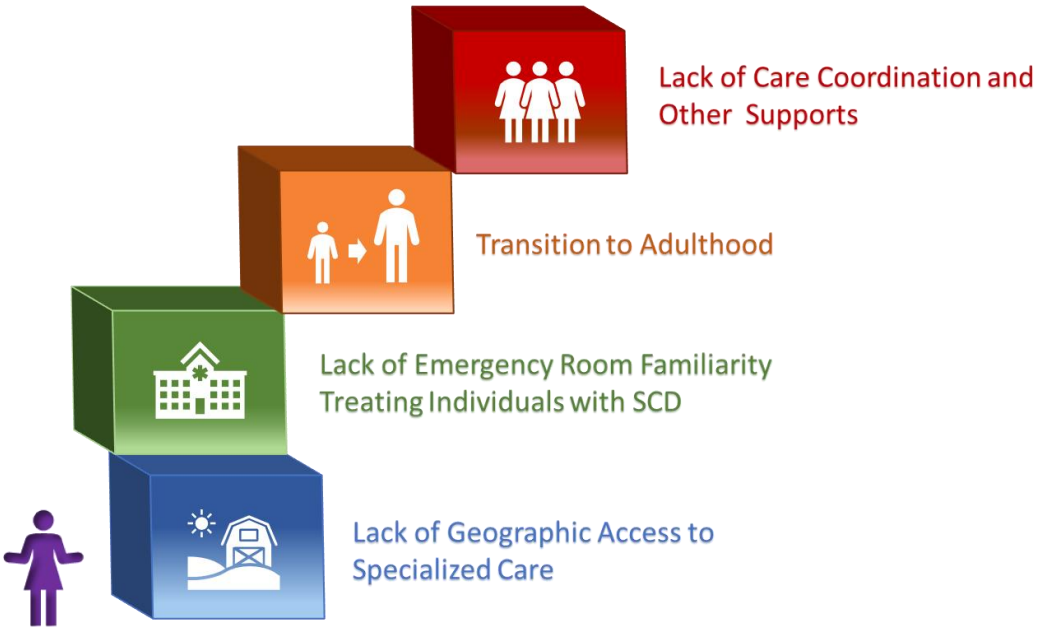
Emergency room providers unfamiliar with SCD may be reluctant to take aggressive steps to manage the pain of patients in crisis. This can prolong the episode and the patient's level of distress.

Adolescents approaching adulthood may be confronted with the need to change providers, if their current provider restricts his or her practice to pediatric patients. SoonerCare members also face a change in benefits when they reach age 19, including a limit on monthly prescription medications and specialist visits, absent prior authorization.

As a life-long chronic condition, SCD also requires a comprehensive approach to care. Patients and families with social stresses and needs may be ill-equipped to manage day-to-day care needs without additional supports.

All of these barriers, alone or in combination, can exacerbate a patient's condition, while also placing strains on the family (Exhibit B-4).

Exhibit B-4 – Examples of Health Disparities/Barriers to Care



SoonerCare Delivery System and Care Management

Delivery System

Individuals with SCD often require support from multiple specialties, with Hematology typically serving as the nexus for their care. The SoonerCare program is open to all licensed and qualified physicians in the State.

Oklahoma has two recognized "centers of excellence" for treatment of persons with SCD, both of which serve SoonerCare members. They are the Jimmy Everest Center at Oklahoma University (OU) Children's Hospital in Oklahoma City and the pediatric Hematology program at Saint Francis Health System in Tulsa.

The OU program was created in 1993 and has served approximately 200 patients over the past two years, over 80 percent of whom were covered through SoonerCare. The program is interdisciplinary and includes Hematologists, behavioral health professionals, a pharmacy liaison, a pediatric nurse practitioner and a nurse coordinator/care manager.

Services include, but are not limited to, a dedicated infusion unit, in-house pharmacy that stocks all newly-approved FDA medications and a bone marrow transplant program. Center staff provide support at the pediatric emergency room and to hospital inpatients.

Jimmy Everest providers care for SCD patients until age 21. The center assists with transitioning the patients to adult care as early as age 13; adults go either to the OU Cancer Center or a local provider that sees adults.

The OU program also is part of a multi-state provider consortium headed by Washington University (St. Louis, MO) that meets monthly to review emerging trends and best practices. The consortium receives funding to support its activities from the federal Health Resources and Services Administration (HRSA). OU is part of the HRSA SCD Southwest Region.

The Saint Francis program, while smaller, serves 90 patients at any point in time. The program is located within the Hematology/Oncology department and includes five physicians and a nurse coordinator. Program Hematologists also support Saint Francis emergency room physicians, as needed.

Supporters of Families with Sickle Cell Disease

Supporters of Families with Sickle Cell Disease is a comprehensive community-based organization serving individuals and families living with sickle cell and thalassemia disease and trait in Oklahoma. The organization is based in Tulsa but works on behalf of families throughout the State.

Supporters of Families with Sickle Cell Disease receives funding through the HRSA grant for the Southwest Region. The organization also has a contract with the OHCA.

The OHCA contract outlines three major goals: Improve quality of life, creating successful working and living interaction for those with Sickle Cell Diseases and Traits; improve health outcomes related to Sickle Cell Disease; and realize cost savings through outreach and education efforts targeting Sickle Cell Diseases and Traits.

The organization's scope-of-work for the OHCA includes[7]:

- Identifying and educating the Oklahoma Sickle Cell Community individuals eligible for Medicaid medical assistance and carriers of Sickle Cell Diseases and Traits;
- Collaborating with OHCA Chronic Care Unit on members with Sickle Cell Disease needing additional community-based supports;
- Identifying and contacting mothers of babies to five (5) years of age and children ages six (6) to eighteen (18) who are newly diagnosed or currently have sickle cell disease in Oklahoma;
- Assembling and distributing Care Kits that provide educational, parental and/or self-care best practices materials for the children identified as newly diagnosed or currently diagnosed with Sickle Cell Disease. Providing education to parent/child on items included in the Care Kit;
- Coordinating statewide collaborative efforts with key organizations in order to identify current resources: current Sickle Cell research and any other key Sickle Cell Entities (national and/or local);
- Creating a free-standing website that is full of resources and an interactive source for Sickle Cell Disease individuals, carriers and their families;
- Enhancing social media presence based on target population, using data and analytics to guide work in this area: Facebook, Twitter, Instagram, Snapchat, and Constant Contact Email Newsletter;
- Pursuing a strong relationship with state agencies for Sickle Cell Disease-Hematology for further community reach: Determining existing and current outreach; determining how this outreach can be enriched and further developed; coordinating and developing a Sickle Cell Disease Outreach Plan and targeting Cell Disease audiences, including members and providers with a youth component for member outreach;
- Training providers regarding the Sickle Cell Disease Outreach Plan;
- Working with clinicians to educate on industry best practices;
- Providing SCD collaboration between patient, families, clinicians; and
- Focusing on compliance critical to medication and treatment plans.

[7] Contract scope-of-work condensed from original language for space and readability. See "Sickle Cell Disease Consulting Contract (Purchase Order 8079004242) Section B.5 for complete language.

Care Management – Current Models

The majority of Medicaid members in Oklahoma are enrolled in SoonerCare Choice, the OHCA's primary care case management model. The program is voluntary and open to most members, the major exceptions being those dually-eligible for Medicare and Medicaid and those receiving long term care services.

SoonerCare Choice enrollees select a primary care provider to serve as their patient centered medical home, or PCMH. The PCMH is responsible for coordinating the member's care needs, including specialist referrals. Members who require a consultation with a specialist generally first must obtain a referral from their PCMH.

The OHCA also has internal programs and contracts with outside organizations to provide enhanced care management to members with complex/chronic conditions such as SCD. These include: the OHCA Chronic Care Unit (CCU), SoonerCare Health Management Program (HMP) and SoonerCare Health Access Networks (HANs) (Exhibit B-5).

Exhibit B-5– SoonerCare Care Management Models

The SoonerCare CCU is located within the OHCA and is staffed by nurses who provide telephonic care management to enrolled members (enrollment is voluntary). The CCU routinely analyzes paid claims data to identify members at highest risk for adverse health outcomes and invites these members (or parents/caregivers of the members) to participate. The criteria for enrollment are: $100,000 in paid claims during the prior year

or $50,000 in paid claims plus five emergency room visits. The CCU provides care management to between 20 and 30 members with SCD at any point in time.

CCU nurses assist with clinical needs, including prior authorizations, transportation and specialist appointments. Nurses inquire about social service needs (e.g., housing or food insecurity) at time of enrollment and make referrals as appropriate. Nurses also reach out to members or parents/caregivers prior to the member's nineteenth birthday, to facilitate the transition from pediatric to adult coverage.

The SoonerCare HMP is a vendor-operated care management program that provides a mix of in-person and telephonic care management to SoonerCare Choice members with complex/chronic health conditions. The HMP is holistic and does not target specific health conditions. The program serves approximately 6,000 participants per year, a small number of whom have SCD.

The SoonerCare HANs are non-profit, administrative entities that work with affiliated providers to coordinate and improve the quality of care provided to SoonerCare Choice members. The HANs employ care managers to provide telephonic and in-person care management to members with complex health care needs who are enrolled with affiliated PCMH providers.

The OHCA contracts with three HANs: University of Oklahoma Sooner HAN; Partnership for Healthy Central Communities HAN; and Oklahoma State University HAN. The HANs' combined enrollment exceeds 300,000, of which approximately 4,000 receive care management over the course of a year.

The HANs historically have provided care management to a small number of members with SCD, identified through data analytics or physician referral. The largest of the three, OU SoonerHAN is preparing to implement a targeted care management program for its members with SCD.

A small number of members with SCD are eligible for long term care and enrolled in one of the OHCA's home- and community-based "waiver" programs[8] or receive care in an institutional setting. These members receive care management as a component of their long term care eligibility.

[8] The term "waiver" refers to the authority under which the home- and community-based services (HCBS) programs operate. States must obtain a waiver of traditional Medicaid rules that cover long term care only in an institutional setting. The largest Medicaid HCBS waiver is the state's ADvantage program for frail elders and adults with physical disabilities.

Care Management – Transition to SoonerSelect

In November 2022, the OHCA released a Request for Proposals (RFP) to contract with risk-based managed care entities (MCEs) to enroll and serve the SoonerCare Choice population, excluding persons eligible due to aged, blind or disabled (ABD) status. The program is to be known as SoonerSelect and contracts are scheduled to take effect in April 2024.

The SoonerSelect MCEs will be responsible for identifying members who would benefit from care management and offering assistance with clinical and social service needs. The SoonerSelect RFP identifies SCD as a priority condition by crafting a case study of an adult member with SCD who is experiencing a pain crisis and asking RFP respondents to describe how they would help the member.

2. SB 1467 – Study Scope and Methods

Senate Bill 1467

During the 2022 regular session, the Oklahoma Legislature enacted Senate Bill (SB) 1467, which was signed into law by the Governor on May 2, 2022. Section 1A of SB 1467 directed the OHCA to:

> *"… conduct an annual review of all medications and forms of treatment for sickle cell disease and services for enrollees with a diagnosis of sickle cell disease. The purpose of the annual review is to determine if the available covered medications, treatments, and services are adequate to meet the needs of enrollees with a diagnosis of sickle cell disease, and whether the Authority should seek to add or recommend additional medications, treatments, or services."*

Section 1B of SB 1467 provided additional direction for the evaluation scope and methods. Specifically, the evaluation should examine:

1. *The extent to which healthcare transitional programs covered under the state Medicaid program prepare, transfer, and integrate emerging adults into the adult care setting from a pediatric setting;*

2. *The extent to which emergency department providers are adequately trained and otherwise prepared to treat and manage sickle cell patients presenting with vaso-occlusive crises including but not limited to the extent to which providers follow clinically validated algorithms and protocols regarding such treatment and management;*

3. *The extent to which sickle cell patients covered under the state Medicaid program are entitled to receive the same standard of care when referred or transferred to an out-of-state facility, and the extent to which the state reimburses such patients for reasonable interstate travel costs; and*

4. *Any additional areas identified by the Authority that impact the care and treatment of individuals in this state living with sickle cell disease or sickle cell trait.*

Sections 1C and 1D provided instructions for collection of information to support the study. Specifically:

> *"When conducting the annual review required by this section, the Authority shall solicit and consider input from the general public, with specific emphasis on seeking input from persons or groups with knowledge and experience in the area of sickle cell disease treatment." (Section 1C)*

> *"To the extent practicable, the Authority shall utilize the Oklahoma State Health Information Network and Exchange created under Section 1-133 of Title 63 of the Oklahoma Statutes to collect information for the purpose of implementing this section." (Section 1D)*

Study Scope and Methods

The OHCA retained the Pacific Health Policy Group (PHPG) to conduct an independent evaluation in accordance with SB 1467 requirements. PHPG is a national consulting firm that specializes in development and evaluation of programs to serve Medicaid populations with special needs. PHPG serves as evaluator of the broader SoonerCare Section 1115 Demonstration under which most Medicaid beneficiaries with SCD receive care.

PHPG organized the study scope around the Legislature's specific areas of inquiry. The evaluation methods similarly were selected to obtain the data necessary to inform findings and recommendations across the areas defined in SB 1467.

The study included six data collection methods:

1. Literature review
2. Provider, care manager and stakeholder interviews
3. Member interviews (structured survey)
4. Analysis of Medicaid eligibility and paid claims data
5. Review of OHCA Drug Utilization Review (DUR) Board activities
6. Review of OHCA coverage policies and managed care strategy

Literature Review

PHPG conducted a broad review of recent national studies and evidence-based guidelines related to care and support for individuals and families with SCD. Sources included federal agencies such as the Centers for Disease Control and Prevention (CDC), National Heart, Lung and Blood Institute (a component of the National Institutes of Health), and private organizations/research institutes such as the American Society of Hematology (ASH), the American College of Emergency Physicians and the National Academy of Sciences and

organizations. PHPG also reviewed web-based materials from organizations dedicated to the SCD community, both in Oklahoma (Supporters of Families with Sickle Cell Disease) and with a national audience (e.g., Sickle Cell Disease Association of America.)

The literature review provided important baseline information and insights into emerging best practices for serving individuals with SCD. Appendix 1 contains a partial listing of reference materials, as well as website links, for interested readers.

Provider and Stakeholder Interviews

PHPG conducted interviews with provider representatives at the two Oklahoma centers-of-excellence for persons with SCD: the Jimmy Everest Center at Oklahoma University Children's Hospital in Oklahoma City and the pediatric Hematology program at Saint Francis Health System in Tulsa. PHPG also contacted representatives at the Oklahoma University's SoonerCare Health Access Network, known as SoonerHAN, which recently established a program specifically for members with SCD.

PHPG interviewed the director of Supporters of Families with Sickle Cell Disease, covering the topics outlined in SB 1467. PHPG and the director also explored opportunities for improving knowledge of SCD within the broader (non-specialist) provider community and enhancing supports for SoonerCare members with SCD. (In addition to the interview, PHPG reviewed a sampling of monthly activity reports submitted by the organization to the OHCA as part of contracting monitoring requirements.)

PHPG also interviewed staff within the SoonerCare Chronic Care Unit (CCU) responsible for providing telephonic care management to CCU-enrolled members with SCD. The interview addressed the topics covered in SB 1467 and the CCU's activities with respect to supporting SCD members with significant medical and social needs.

In addition, PHPG participated as an observer in a virtual Town Hall Meeting organized by Supporters of Families with Sickle Cell Disease in collaboration with the Oklahoma State Department of Health (OSDH). The meeting took place on November 30, 2022 and was open to the public via the internet. Attendees were encouraged to share their perspectives on access to care, service gaps, Medicaid coverage, community supports, pain management and evidence-based care. Appendix 2 includes a copy of the meeting invitation.

Supporters of Families with Sickle Cell Disease also shared with PHPG the results of a survey of 20 primary care and specialty providers the organization conducted in April 2022, in collaboration with OSDH. Findings from the survey are noted throughout the report.

Member Interviews

PHPG developed a structured member survey instrument to be administered by telephone. The survey addressed all of the areas of inquiry specified in SB 1467. Appendix 3 contains a copy of the survey instrument.

PHPG mailed advance letters to every household with a SoonerCare member with SCD before placing calls. Surveyors made up to six contact attempts over a 30-day period, including in the evening and on weekends. (Members also were provided a toll-free number to call at any time to participate in the survey.)

Sixty-seven surveys were completed with adult members or the parents/caregivers of members under age 18[9]. The findings were analyzed both at a statewide level and separately for respondents living in metropolitan and rural areas[10].

Analysis of Eligibility Files and Paid Claims

As an evaluator of the broader SoonerCare program, PHPG has direct access to Medicaid eligibility and claims systems. PHPG produced an eligibility and claims extract for State Fiscal Years 2020 – 2022, to develop a multi-year eligibility and utilization profile of SoonerCare members with SCD.

The analysis examined:

- Place of residence for members with SCD
- Service utilization patterns, including primary care, specialty care, prescription drugs, emergency room visits and hospital admissions
- Per member per month expenditures

Note: The three-year period overlapped with the COVID-19 Public Health Emergency (PHE), which disrupted care patterns for all populations. Trend lines should be viewed with caution, given the impact of COVID-19 on the health care system.

[9] Supporters of Families with Sickle Cell Disease and OSDH also surveyed individuals with SCD concurrent with the provider survey (SoonerCare and other). The survey instrument overlapped in part with PHPG's and results for overlapping areas of inquiry were similar across the two surveys. Both surveys also explored unique areas of interest.

[10] The great majority of respondents resided in metropolitan areas of the State, consistent with the SCD population's overall distribution. Results are presented in the report at the statewide level only.

Review of DUR Board Activities

Medicaid covers all FDA-approved medications, although prescription coverage can be subject to limits (e.g., dosage limits or a limit to the number of scripts per month for adults) and prior authorization requirements. SoonerCare's prescription drug benefit is overseen by the OHCA Drug Utilization Review Board, whose mission is to advise the OHCA about the appropriate and optimal use of pharmaceuticals for Oklahoma Medicaid beneficiaries.

The DUR Board meets monthly and includes independent physicians and pharmacists, as well as OHCA representatives. The Board devotes a portion of one meeting each year to review advances in treatment of anemia-related disorders, including SCD. The most recent such meeting was held in October 2022.

PHPG examined DUR Board materials from the October 2022 meeting, to document updates in policy with respect to treatment of SCD. PHPG also interviewed OHCA DUR staff regarding SCD-related activities.

Review of OHCA Managed Care Strategy

The OHCA is implementing a managed care strategy under which a majority of the SoonerCare population – including most members with SCD – will be enrolled into Managed Care Entities (MCEs) responsible for service delivery and care coordination. MCE enrollment is scheduled for April 2024.

PHPG reviewed the MCE Request for Proposals (RFP) and related materials to document the OHCA's proposed approach to serving members with complex/chronic conditions under the MCE model.

Summary of Evaluation Scope and Methods

Exhibit B-6 below cross-references the evaluation scope and corresponding data collection methods.

Exhibit B-6 – Evaluation Scope and Methods

Evaluation Component	Eligibility/ Claims	DUR Reports	OHCA Policy	National Research	Stakeholder Interviews			
					Members/ Families	Providers	CCU/HAN	Other
1. Do SoonerCare members with SCD have access to all necessary services?								
Utilization and expenditure trends	✓	✓						
Availability of recommended services (including PCMH awareness of SCD treatments)		✓	✓	✓	✓	✓	✓	✓
Out-of-state referrals			✓		✓	✓	✓	✓
2. Do Oklahoma emergency room providers have the appropriate training and resources to care for members in crisis?								
Training and resources	✓			✓	✓	✓	✓	✓
Adherence to algorithms/protocols				✓		✓	✓	✓
3. Do SoonerCare members with SCD have appropriate supports to navigate the health care system?								
Acute case management			✓		✓	✓	✓	✓
Chronic care management			✓		✓	✓	✓	✓
Transition to adulthood			✓		✓	✓	✓	✓
4. How can the program be strengthened?								
Member access and supports					✓		✓	✓
Provider access and supports						✓	✓	✓

Report Organization

Section C begins by providing additional data on the characteristics of SoonerCare members with SCD. It then presents evaluation findings for the areas of inquiry outlined in SB 1467.

Section D presents summary recommendations for enhancing care and supports for persons with SCD and their families. All recommendations are intended to be actionable in in the next 12 to 18 months.

Report appendices are included after Section D.

C. EVALUATION FINDINGS

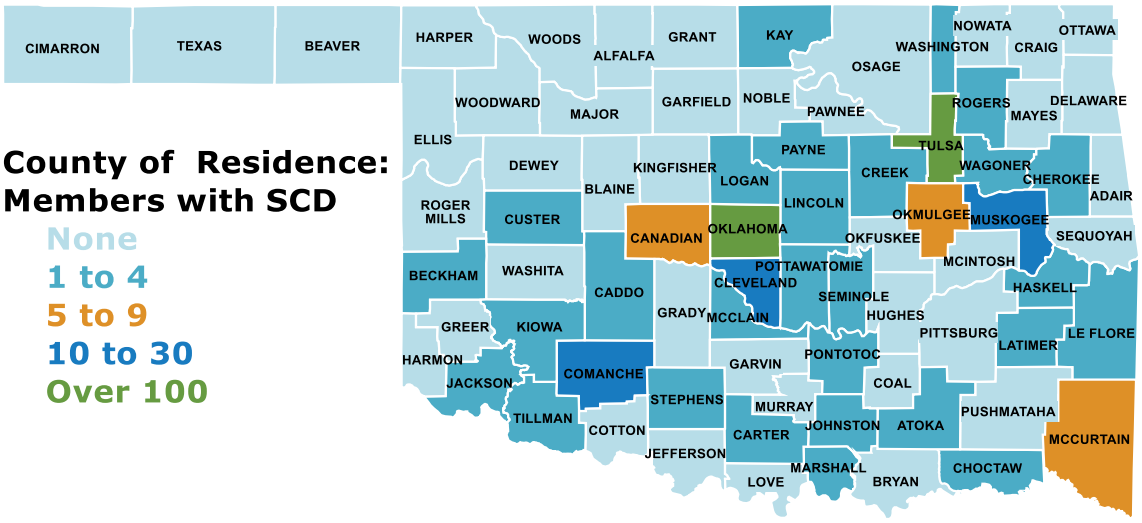
1. *Characteristics of SoonerCare Members with SCD*

PHPG analyzed eligibility data for State Fiscal Year (SFY) 2022 (July 2021 – June 2022), to profile the demographic characteristics of SoonerCare members with SCD, including places of residence, age ranges and SoonerCare aid categories.

County of Residence

SoonerCare members with SCD are not evenly distributed throughout the State. Most reside in Oklahoma and Tulsa Counties, each of which is home to over 100 members with SCD (Exhibit C-1). The next most populated counties are Cleveland, Comanche and Muskogee, each with between 10 and 30 members. There are 31 counties with at least one, but fewer than 10 members with SCD; 41 counties have no members with SCD[11].

Exhibit C-1 – SoonerCare Members with SCD by County of Residence



In total, the top five counties account for over 85 percent of all members with SCD (Exhibit C-2 on the following page).

[11] PHPG adhered to OHCA guidelines for identifying members with chronic health conditions. A member was included in the analysis data set for a particular year if she or he had at least two paid claims with a Sickle Cell Disease diagnosis code.

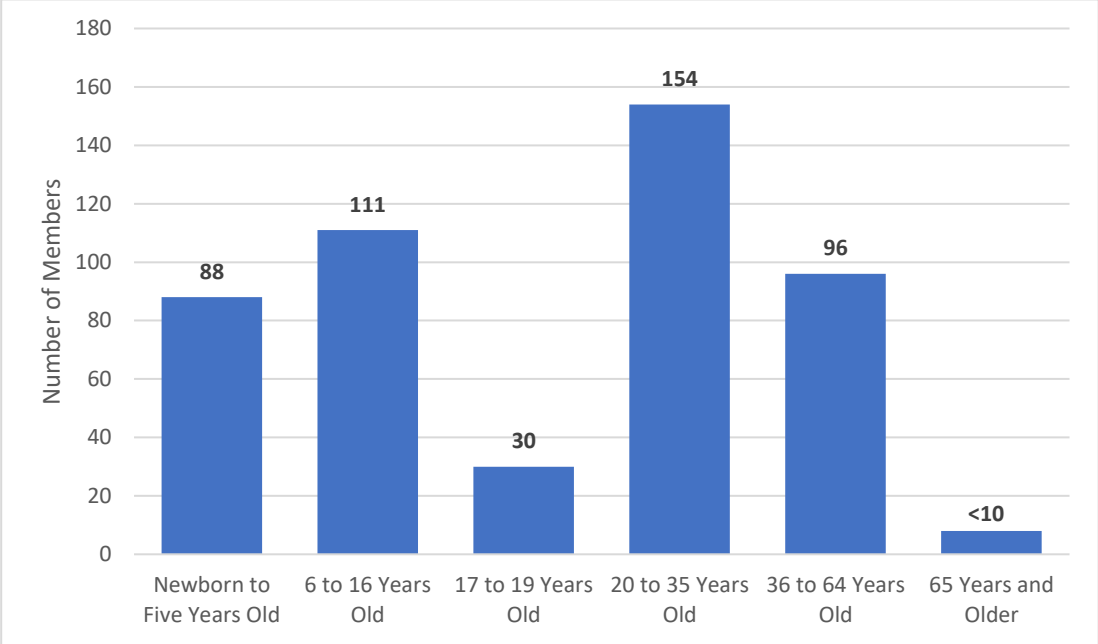
Exhibit C-2 – SoonerCare Members with SCD – Top Five Counties

County	Number of Members	Percent of Total	Cumulative Percentage
Oklahoma	207	42.5%	42.5%
Tulsa	142	29.2%	71.7%
Comanche	28	5.7%	77.4%
Cleveland	20	4.1%	81.5%
Muskogee	19	3.9%	85.4%
Other Counties/Out of State	71	14.6%	100.0%
Total	**487**	**100.0%**	**100.0%**

Age Ranges

Approximately 45 percent of members with SCD in SFY 2022 were under the age of 20; this included 30 older adolescents approaching the transition from child to adult coverage. Most adults were 20 to 35 years in age, although a significant number were in the 36 to 64 age cohort (Exhibit C-3).

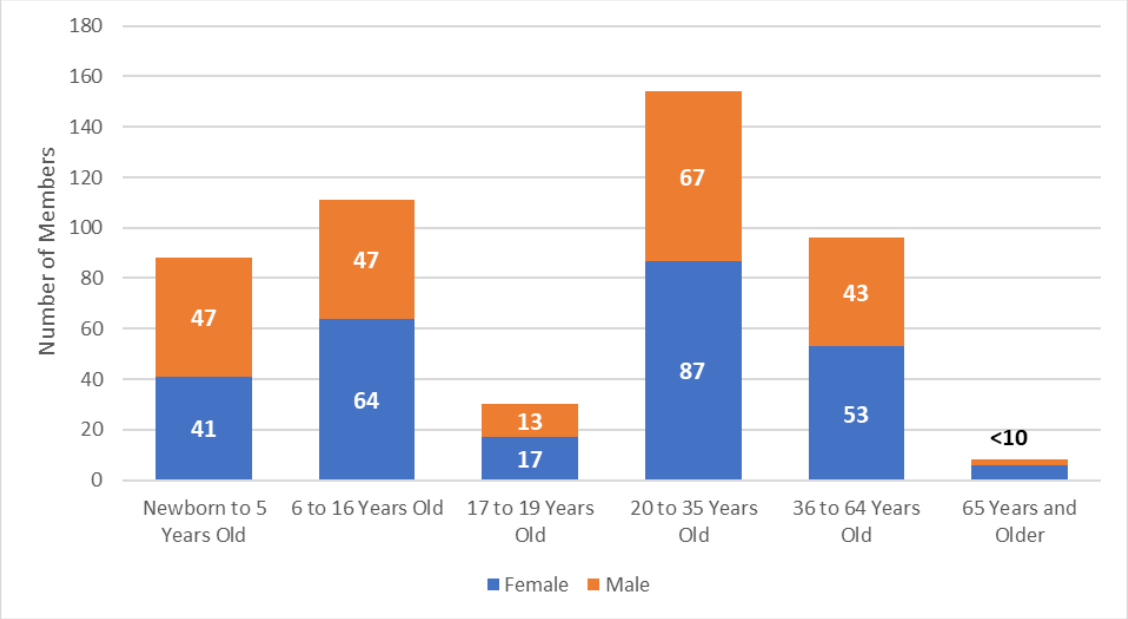
Exhibit C-3 – SoonerCare Members with SCD by Age Range



Age and Gender

Females in SFY 2022 outnumbered males in all age cohorts except newborns to five years old (Exhibit C-4).

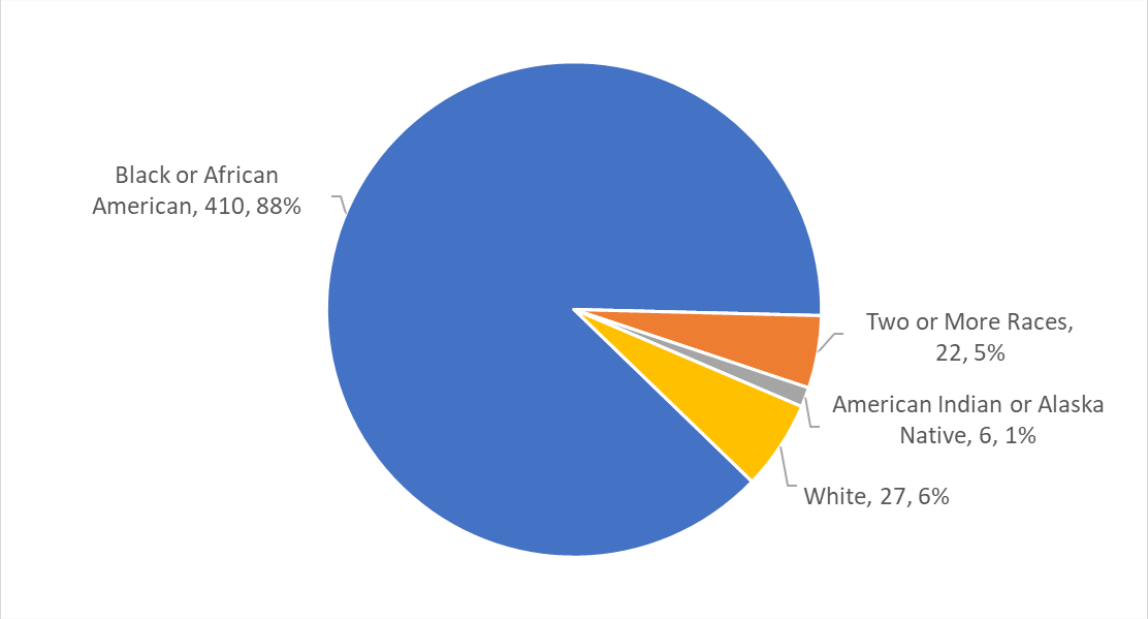
Exhibit C-4 – SoonerCare Members with SCD by Age Range and Gender



Race (Self-Reported)

African Americans comprised nearly 90 percent of members with SCD in SFY 2022, based on self-reported race (Exhibit C-5).

Exhibit C-5 – SoonerCare Members with SCD by Race (Self-Reported)



2. Access to Covered Services

PHPG evaluated the adequacy of SoonerCare covered services for members with SCD through the following methods:

- Medicaid coverage policies
- Paid claims data
- Provider, stakeholder and member interviews

Medicaid Coverage Policies – General

Medicaid programs operate in accordance with Title 42 of US Code, which defines mandatory and optional services, the latter of which can be covered at a state's discretion. Mandatory services include inpatient and outpatient hospital care, physician services and lab/X-ray services, among others. Major optional services include prescription drugs, therapies, vision and dental care. Oklahoma covers all major optional services, as well as most other optional service types[12].

Medicaid coverage for individuals up to age 21 in Oklahoma and all other states is defined further within the Early and Periodic Screening, Diagnosis and Treatment (EPSDT) section of federal law and regulations[13]. Under EPSDT, state Medicaid programs are responsible for, "…providing early and periodic screening and diagnosis of eligible Medicaid beneficiaries under age 21 to ascertain physical and mental defects, and providing treatment to correct or ameliorate defects and chronic conditions found." The treatment requirement encompasses all medically necessary services, even if not otherwise covered under the state's Medicaid program.

Unlike for members under age 21, states can impose limits on covered services for members 21 and older. Oklahoma limits the number of covered prescriptions to six per month and specialist physician visits to four per month, among other limitations. The OHCA has a prior authorization process that can be used to exceed monthly limits when medically necessary.

[12] See Mandatory & Optional Medicaid Benefits | Medicaid for a complete listing of federally-defined mandatory and optional services.
[13] See eCFR :: 42 CFR Part 441 Subpart B -- Early and Periodic Screening, Diagnosis, and Treatment (EPSDT) of Individuals Under Age 21

Medicaid Coverage Policies – Out-of-State Services

The OHCA covers out-of-State services for SoonerCare members in accordance with State and Federal law[14]. In 2019 the Oklahoma Legislature passed HB 2341, which limited SoonerCare members' services to in-State providers when possible[15].

In accordance with the statute, the OHCA modified its policies to require prior authorization for out-of-State services, other than care rendered to members living near the State border and seeing a contracted provider within 50 miles of the border.

Providers must submit a prior authorization request form, for review by OHCA staff dedicated to this function. The form must be accompanied by:

- Documentation to establish the medical necessity of services requested, such as medical records
- Letter of medical necessity or other thorough summary document that includes:
    - Summary of the member's condition and history of treatment related to the request
    - History of other providers who have evaluated, treated or consulted member related to the request
    - Recommended treatment or further diagnostic needed
    - Why medical care cannot be completed in Oklahoma or the next closest location

Appendix 4 includes a copy of the prior authorization form and the OHCA notice to members and providers explaining the agency's updated policy.

(Note: PHPG reviewed with the OHCA requests for out-of-State care. Due to the low volume of such requests, PHPG is not presenting data in the report to ensure no violations of HIPAA privacy rights.)

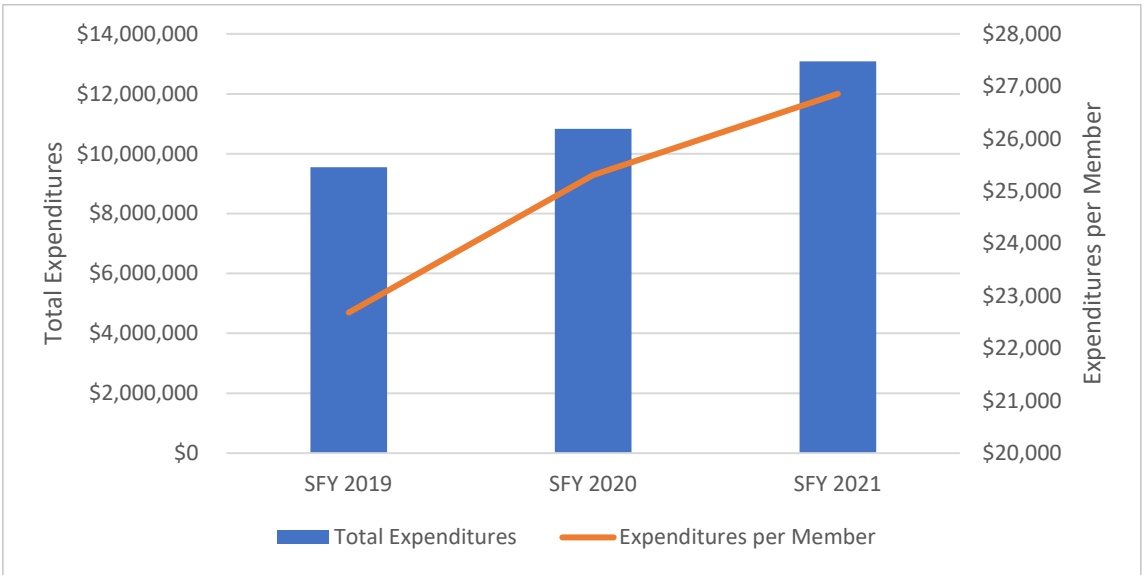
[14] Includes transportation and lodging costs, as applicable.

[15] Section 59.A: Where practicable and in accordance with state and federal law, the state Medicaid program shall not contract with an out-of-state medical provider for treatment that is available from one or more providers licensed and practicing in the State of Oklahoma.

Paid Claims Analysis

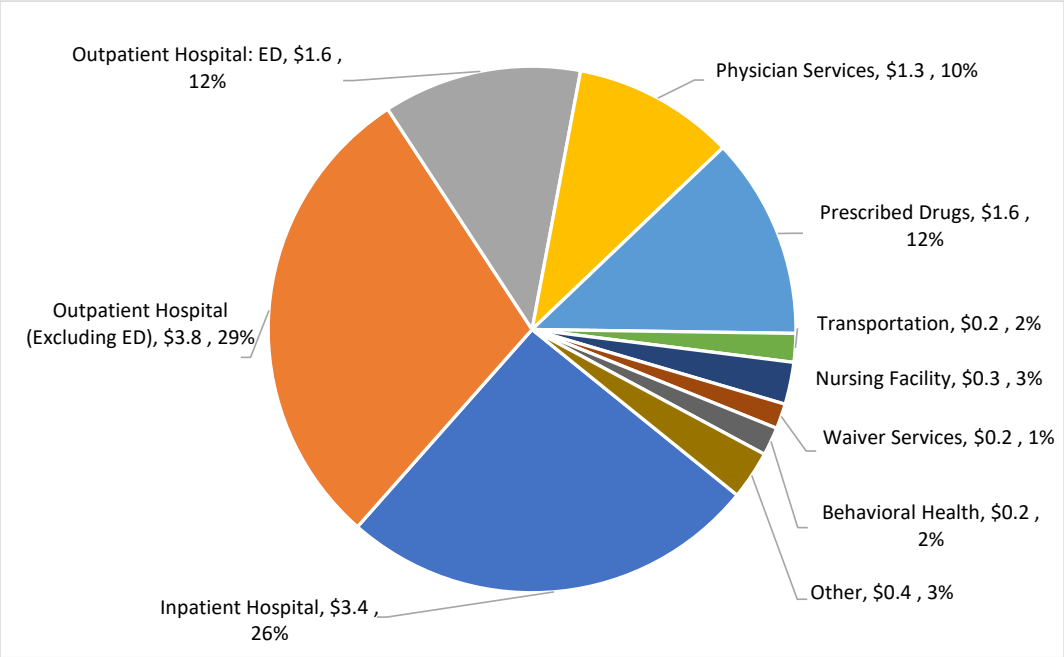
Paid claims for SoonerCare members with SCD totaled approximately $13.1 million in SFY 2022, up from $10.8 million in SFY 2021 and $9.5 million in SFY 2020. The increase was due in part to growth in members with SCD and in part to growth in expenditures per member. The average annual expenditure per member in SFY 2022 was $26,861, up from $25,304 in SFY 2021 and $22,680 in SFY 2020 (Exhibit C-6).

Exhibit C-6 – Expenditure Trend – SFY 2020 to SFY 2022



The largest service category, in terms of paid claims, was outpatient hospital, excluding emergency room visits (Exhibit C-7 on the following page).

Exhibit C-7 – Expenditures by Service Category (SFY 2022)



Although the average expenditure per member in 2022 was $26,861, there was a wide range between low- and high-cost members. The top six percent of members averaged over $190,000 each and accounted for approximately 46 percent of total expenditures. Conversely, the bottom 31 percent of members accounted for less than two percent of total expenditures (Exhibit C-8 below and on the following page).

Exhibit C-8 – Expenditures per Member by Expenditure Range (SFY 2022)

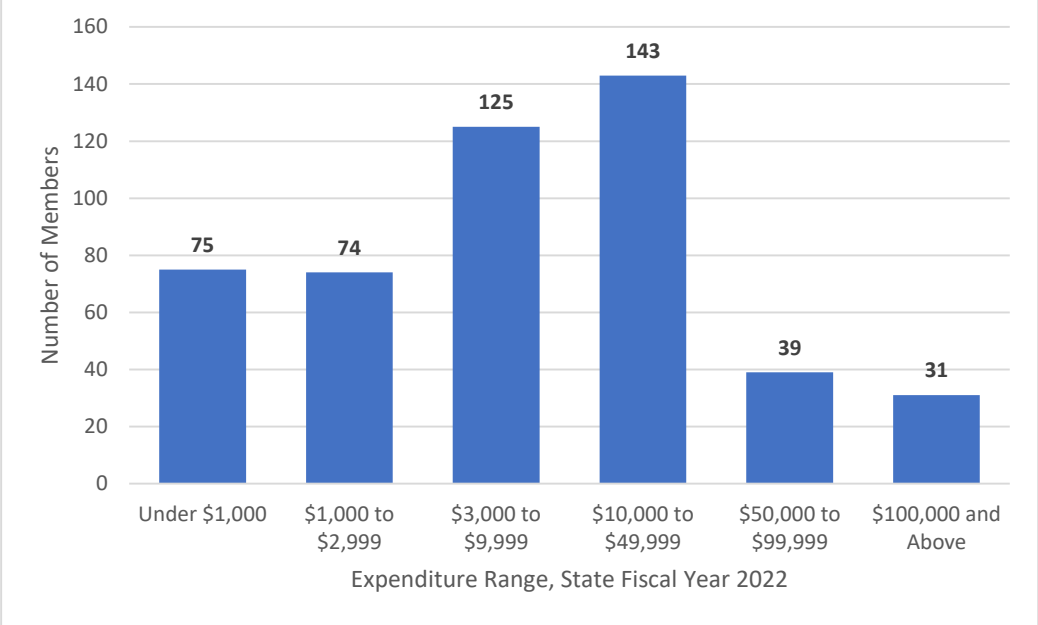


Exhibit C-8 – Expenditures per Member by Expenditure Range (SFY 2022) cont'd.

Expenditure Range	Number of Members	Percent of Members	Average Per Member	Percent of Expenditures
Under $1,000	75	15.4%	$483	0.3%
$1,000 to $2,999	74	15.2%	$1,963	1.1%
$3,000 to $9,999	125	25.7%	$5,730	5.5%
$10,000 to $49,999	143	29.4%	$23,617	25.8%
$50,000 to $99,999	39	8.0%	$70,812	21.1%
$100,000 and above	31	6.4%	$194,987	46.2%
Total	**487**	**100.0%**	**$26,861**	**100.0%**

The majority of members with SCD saw a SoonerCare Choice PCMH provider at least once in SFY 2022. Members seeing the two most common provider types, Family Practitioners and Internists, averaged approximately six visits per year (Exhibit C-9)[16].

Exhibit C-9 – PCMH Provider Type Activity (SFY 2022)

Primary Care Provider Type	Number of Members	Number of Visits	Average Per Member	Percent of Members
Family Practitioner	148	667	4.5	30.4%
General Pediatrician	89	253	2.8	18.3%
Internist	121	986	8.1	24.8%
General Practitioner	30	82	2.7	6.2%

Members saw a variety of specialists, the most common of which were Hematology/Oncology providers. The Hematology/Oncology providers treating members with SCD are concentrated in Oklahoma City and Tulsa, with smaller numbers practicing elsewhere, including in Lawton and Muskogee.

[16] Physician Assistants not shown on table. Data is not unduplicated by member.

Nearly 400 members with SCD had at least inpatient stay during the SFY 2020 – SFY 2022 period; most of these members (263) had multiple admissions (Exhibit C-10). In nearly 69 percent of the admissions, the primary diagnosis was for treatment of Sickle Cell Anemia (60.4 percent), Sickle Hemoglobin (4.7 percent) or Thalassemia (1.3 percent). This is consistent with the need for persons with SCD to be hospitalized for treatment of acute pain crises, among other complications. (Appendix 5 contains a complete listing of admitting diagnoses during the three-year period.)

Exhibit C-10 – Inpatient Hospital Activity (SFY 2020 – 2022)

Admissions per Member	Number of Members	Total Admissions	Average Per Member	Percent of Admissions
1 Admit	134	134	1.0	7.0%
2 – 3 Admits	127	311	2.4	16.4%
4 – 6 Admits	61	301	4.9	15.9%
7 – 10 Admits	39	339	8.7	17.9%
Over 10 Admits	36	811	22.5	42.8%
Total	**397**	**1,896**	**4.8**	**100.0%**

Provider Interviews

Providers at Oklahoma's two SCD centers-of-excellence did not identify any coverage gaps, strictly from an insurance perspective, that presented barriers to their treatment of patients with SCD. This was unsurprising, given that Medicaid coverage generally is comparable to commercial insurance in scope.

However, Jimmy Everest providers mentioned that arranging transportation can be a burden, particularly for members traveling long distances, and both centers noted that the requirement for a PCMH specialist referral can delay care. The referral issue occurs when the centers, which do not operate as PCMH sites, determine there is a need for the member to be referred to another specialty (e.g., a nephrologist or neurologist).

The centers cannot make the referral directly but instead must direct patients back to their PCMH for the authorization. (The same restriction applies to any other specialist wishing to make a referral to another specialist.) While this ensures the PCMH is kept informed of the patient's total course of care, it is exceedingly rare (according to the centers) for the PCMH to do other than simply approve the referral as requested.

In some managed care systems, the requirement to go back to the PCMH is not imposed on specialist providers with a proven record of making appropriate referrals. Specialists still must share any referral results with the PCMH, a step that is becoming easier as health systems move to electronic health records and states implement health

information exchanges (HIEs). (The OHCA is in the process of implementing Oklahoma's HIE, known as OK SHINE.)

Member Interviews

PHPG's member survey included questions about the member's experience accessing non-emergency care within the SoonerCare program. (Emergency care was asked about separately; results are presented in the next section of the report).

All respondents reported having a PCMH provider, usually a Family/Nurse Practitioner or Pediatrician. Seventy five percent reported also seeing a specific specialist physician on a regular basis; for most, this was a Hematologist/Oncologist, although other specialties mentioned included Cardiology, Neurology and Pulmonology.

Twenty-seven percent reported having a care manager. Although this was a minority of respondents, by comparison less than two percent of the total SoonerCare Choice population receives care management through one of the previously-described OHCA programs. Nevertheless, nearly three-in-four respondents did not have a care manager.

Respondents next were asked whether they had received age-appropriate preventive services recommended for persons with SCD, as well as any of the evidence-based treatments considered to be part of the standard of care (depending on a patient's particular condition and needs).

Exhibit C-11 below summarizes the responses by service, in descending order of "Yes" responses. The percentage answering "Yes" to the preventive services ranged from approximately 94 percent for blood pressure checks down to approximately 42 percent for pneumococcal vaccine. The "Yes" rate was below 80 percent for six of the nine recommended preventive services.

Approximately 75 percent reported having a treatment plan for pain control, although these are not necessarily specific to crisis episodes or emergency room care. Smaller percentages reported receiving blood transfusions or chelation therapy.

Exhibit C-11 – Member Survey – Service Use

Question – *"Please tell me if you (your child) receive…"*	Yes	No	Not Sure
Preventive Care			
1. Had blood pressure checked at least once in the past twelve months (if three years or older)	94.3%	5.7%	---
2. Had lungs checked at least once in the past twelve months	83.9%	14.5%	1.6%

Question – *"Please tell me if you (your child) receive…"*	Yes	No	Not Sure
3. Had heart checked at least once in the past twelve months	80.6%	16.1%	3.2%
4. Receive daily penicillin (if younger than five years)	70.6%	29.4%	---
5. Had kidneys checked at least once in the past twelve months (if twelve months or older)	60.0%	26.7%	13.3%
6. Had a head X-ray called a 'transcranial doppler' at least once in the past twelve months	58.6%	37.9%	3.4%
7. Had retinas (eyes) examined at least once in the past twelve months (if ten years or older)	54.3%	37.1%	8.6%
8. Receive Hydroxyurea medication (if older than nine months)	50.0%	48.3%	1.7%
9. Received a pneumococcal vaccine (shot to prevent pneumonia) in the past twelve months	41.9%	43.5%	14.5%
Treatments			
10. Have a treatment plan to control pain	75.8%	24.2%	---
11. Received one or more blood transfusions in the past twelve months	38.7%	61.3%	---
12. Currently receive chelation therapy for iron overload	3.2%	93.5%	3.2%

Respondents next were asked to rate their satisfaction with aspects of the care they or their child received from their regular doctor and other members of the care team for treatment of their Sickle Cell Disease. Exhibit C-12 below presents results in the order asked.

Over 90 percent of respondents were "very" or "somewhat" satisfied with each of the aspects of care, the exception being "finding doctors who know how to care for patients with Sickle Cell Disease". A portion of respondents were "somewhat" or "very" dissatisfied with each of the items.

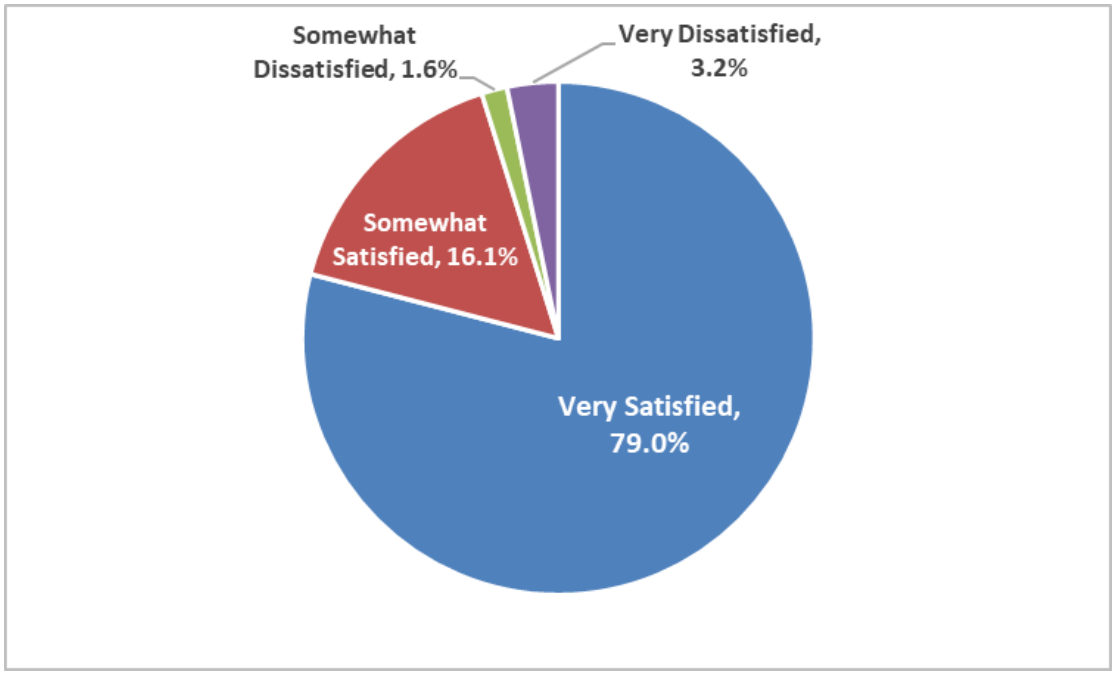
Exhibit C-12 – Member Satisfaction with Aspects of Care

Aspect of Care	Very Satisfied	Somewhat Satisfied	Somewhat Dissatisfied	Very Dissatisfied	Not Sure
1. Finding doctors who know how to care for patients with Sickle Cell Disease	77.4%	8.1%	9.7%	4.8%	---
2. Being able to schedule doctor's appointments when you need them for treatment of your (your child's) Sickle Cell Disease	74.2%	17.7%	3.2%	4.8%	---
3. Getting the right services and treatments, other than medication,	87.1%	6.5%	1.6%	4.8%	---

Aspect of Care	Very Satisfied	Somewhat Satisfied	Somewhat Dissatisfied	Very Dissatisfied	Not Sure
for your (your child's) Sickle Cell Disease					
4. Getting the right medications for your (your child's) Sickle Cell Disease	88.7%	4.8%	---	4.8%	1.6%
5. Getting extra support and help if you need it to manage your Sickle Cell Disease	83.9%	8.1%	3.2%	4.8%	---
6. Being listened to and understood when you have concerns about your (your child's) Sickle Cell care in general, other than pain management	85.5%	8.1%	1.6%	4.8%	---
7. Being listened to and understood when you have concerns about managing your (your child's) Sickle Cell pain	87.1%	6.5%	1.6%	4.8%	---

Respondents also were asked to rate their overall satisfaction with the care received from regular doctors and other members of the care team. Exhibit C-13 below groups the responses, which were similar to the ratings for individual aspects of care.

Exhibit C-13 – Overall Satisfaction with Non-Emergency Care



Respondents were asked whether they had ever sought approval to receive care from an out-of-State provider. None had.

Respondents also were invited to describe their experiences with the health care system. They offered a mix of positive and negative comments.

> *"SoonerCare saved my life. Before I was on SoonerCare…I was not getting the care that I needed. Once I found out I could get SoonerCare, I started getting the medical treatment that I needed. I am in less pain now and am working on getting healthier. I also like that now my doctors all talk to each other and can see what each other has done."*
>
> *"I have received better care now than I did before I was living in Oklahoma. My doctors here listen to me and what I want. I hate taking medications and my doctors now offer me other options for my treatment."*
>
> *"All of (my daughter's) doctors make sure (she) is getting everything she needs. I got a call yesterday saying she needs to start taking a supplement because they saw her levels were low. I feel they are on top of her health."*
>
> *(My son's) Hematologist and Oncologist are good. His regular doctor does not know anything about SCD though."*
>
> *"My PCP does the best she can to help me manage my Sickle Cell disease but she is not a specialist. I have trouble managing my pain and I have been on hydrocodone for years now and I do not feel it works anymore, my body is used to it. My PCP does not know what else to prescribe though. There are no specialists in my area (outlying suburb of Oklahoma City) for me to go to."*
>
> *"I cannot get treatment for my Sickle Cell disease. I have been told that I have the disorder by some doctors and others tell me that I only have the trait. I tell them that I am in terrible pain, especially in the winter, and that I do have it, but they don't listen. Even if I have a mild case, I still need treatment for it. My regular doctor does not know anything about it…"*

The providers responding to the Supporters of Families with Sickle Cell Disease/OSDH survey acknowledged the need for better education, both for themselves and patients. A majority said they would welcome the opportunity to learn about current research and new therapies, emerging new medications and treatments and complications that arise

in individuals with SCD. A majority also said they lacked adequate patient-centered educational materials on managing the condition.

Town Hall Meeting

The November 30, 2022 virtual Town Hall meeting was attended by adult members with SCD and parents of children with SCD, as well as representatives from the OHCA, OSDH and the federal Health Resources and Services Administration (HRSA). The meeting was organized and facilitated by Supporters of Families with Sickle Cell Disease. (PHPG and various agency representatives participated as observers only.)

Attendees expressed similar concerns to survey respondents about the lack of knowledge of SCD within the primary care provider community. This can create a potential barrier to care given the requirement given the important role played by PCMH providers in managing the needs of SoonerCare Choice members.

> *"I just moved here from Chicago, and for me it's been pretty difficult. I feel like my doctor, my Hematologist, I feel like him and his team, especially when I am admitted into the hospital, I don't feel like I am being heard and they don't listen to me as far as what I am telling them I need to get better. I have been dealing with this since I was two, I know my body. I know the different things doctors have tried for me. I know what works, what doesn't. I feel like I am just not being heard as a patient so it's been difficult for me especially me being a new patient. It's been difficult for sure. I've only been here a little over a year and all my hospital stays have been hard."*

> *"I am sickle cell warrior. I am 27 years old. I just had my second baby. So far from having my second baby my interactions with the doctor have been different. They have been helpful for the most part. It did take a pretty long time for me to be able to have this type of relationship with my doctor…. before it took a lot of having to advocate for myself with the doctor letting him know like some of what you're doing for my care isn't helping me with feeling better. It took a couple years, it took years, with me and him having this conversation over and over again. So, we're definitely at point we can communicate better. Before he was very dismissive, didn't want to understand and telling me I can find a new doctor if I wanted to. So, it was just very difficult for a couple of years."*

Access to Covered Services – Summary of Findings

PHPG's review of the SoonerCare program's performance with respect to offering access to covered services found:

- SoonerCare provides coverage to the same extent as commercial insurance and other state Medicaid programs for children. The program does impose some service limits for adults age 21 and older but neither providers nor members with SCD identified any of these limits as posing a barrier to care.
- Members generally are satisfied with their care but some expressed dissatisfaction with the knowledge of SCD among PCMH providers. This concern was voiced both by survey respondents and Town Hall participants. It also was reflected in the desire of provider survey respondents to learn more about treatments for SCD.
- The need for PCMH authorization of all specialist visits was identified as a barrier to timely care by the two SCD centers of excellence.

The majority of members with SCD will be enrolled into SoonerSelect MCEs in April 2024. The OHCA already has highlighted the importance of SCD within SoonerSelect by highlighting it within the RFP.

The transition provides an opportunity for educating PCMH providers about evidence-based best practices for care of members with SCD. The OHCA, in conjunction with SoonerSelect MCEs, the two centers-of-excellence and Supporters of Families with SCD, could conduct a coordinated educational campaign targeting PCMH providers in counties with SCD members. The OHCA could explore the potential for offering continuing education credits to participants.

It is common in managed care systems for processes to be put in place to facilitate specialist-to-specialist referrals. For example, specialists that have a proven record of making appropriate referrals are often exempted from the requirement to send patients back to their PCMH to obtain the formal referral authorization. PCMH providers can be kept informed through notification requirements. (The implementation of the HIE also will facilitate information sharing between PCMH providers and specialists in different health care systems.)

The OHCA can encourage SoonerSelect MCEs to put such a policy in place for members with SCD, if a broader policy is not already in effect. The OHCA also could explore modifying its internal policies for the population that will not be enrolled into SoonerSelect.

3. Emergency Room Provider Training and Resources

PHPG evaluated Emergency Room activity and the adequacy of Emergency Room provider training and resources through:

- Medicaid coverage policies
- Paid claims data
- Provider, stakeholder and member interviews

Medicaid Coverage Policies

Oklahoma was one of the first states to be affected severely by the opioid crisis and also one of the first to take concerted action, both legislative (through the State's Anti-Drug Diversion Act) and in policy. The OHCA in the past decade implemented a strategy for lowering gradually the opioid dosage that providers could prescribe most patients without prior authorization. The OHCA also expanded its contract with the Health Management Program vendor to include pain management education for providers with a history of prescribing opioids more extensively than their peers.

One unintended consequence of the campaign to reduce inappropriate use of opioids has been to make it more difficult for members with SCD to receive necessary medication when experiencing a pain crisis. Providers unfamiliar with the nature of the disease can be reluctant to accede to a patient's urgent request for prescription pain medication.

In its 2021 regular session, the Oklahoma Legislature addressed the pain management needs of persons with conditions like SCD through passage of SB 57, which amended the State's Anti-Drug Diversion Act. At the recommendation of the patient advocate community, new language was inserted that states:

> *"Nothing in the Anti-Drug Diversion Act shall be construed to require a practitioner to limit or forcibly taper a patient on opioid therapy. The standard of care requires effective and individualized treatment for each patient as deemed appropriate by the prescribing practitioner without an administrative or codified limit on dose or quantity that is more restrictive than approved by the Food and Drug Administration (FDA)."*[17]

This language removed a statutory barrier to treating members with SCD, and OHCA coverage policy aligns with the statute. However, the experience of members with SCD who seek treatment in an Emergency Room still varies based on the knowledge of

[17] Section 2.K of the Act.

providers about their condition and their recognition (or lack thereof) of the need to prescribe very high doses of pain medication.

Paid Claims Data

SoonerCare members with SCD who experience a pain crisis must be treated in a hospital setting, either in the emergency room or as an inpatient. Medications are administered parenterally and require continuous monitoring.

In SFY 2022, 327 out of 487 members with SCD (67.1 percent) had at least one emergency room visit. The 487 members sought care in the emergency room an average of 4.3 times each; the subset with one or more visits sought care an average of 7.6 times each.

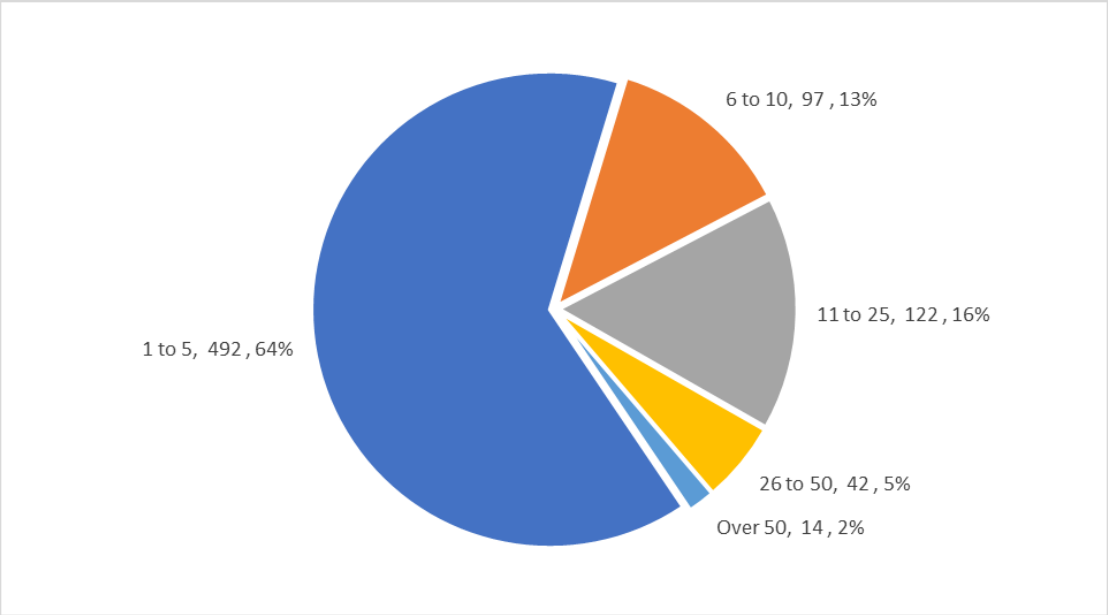
Within the 7.6 average, the range of visits varied widely. The top 26 members accounted for over 50 percent of all emergency room visits (Exhibit C-14).

Exhibit C-14 – Emergency Room Visit Activity (SFY 2022)

Number of Visits	Number of Members	Number of Visits	Average Per Member	Percent of Visits
1 Visit	86	86	1.0	3.5%
2 – 3 Visits	94	224	2.4	9.0%
4 – 8 Visits	96	522	5.4	21.1%
9 – 15 Visits	25	279	11.2	11.3%
16 – 30 Visits	14	294	21.0	11.9%
31 – 50 Visits	7	274	39.1	11.0%
Over 50 Visits	5	796	159.2	32.2%
Total	**327**	**2,475**	**7.6**	**100.0%**

The emergency rooms at OU Health Sciences Center and Saint Francis have evidence-based protocols for treatment of patients in crisis and providers are familiar with how to treat the condition. However, the majority of emergency room physicians from SFY 2020 to 2022 who treated patients with SCD saw, on average, only one or two cases per year (Exhibit C-15 on the following page).

Exhibit C-15 – Emergency Room Physician Activity (SFY 2020 – SFY 2022)



Number of Encounters	Number of Physicians	Percent of Physicians	Total Encounters	Average Per Physician
1 – 5 Encounters	492	64.1%	961	2.0
6 – 10 Encounters	97	12.6%	755	7.8
11 – 25 Encounters	122	15.9%	2,001	16.4
26 – 50 Encounters	42	5.5%	1,498	35.7
Over 50 Encounters	14	1.8%	1,295	92.5
Total	**767**	**100.0%**	**6,510**	**8.5**

Member Interviews

PHPG's member survey included questions about the member's experience accessing care in the emergency room, if applicable. Fifty-eight percent of respondents reporting going to the emergency room in the past 12 months, slightly below the rate for the population overall. The average number of visits across all respondents was 2.1; the average among those who visited the emergency room was 3.6.

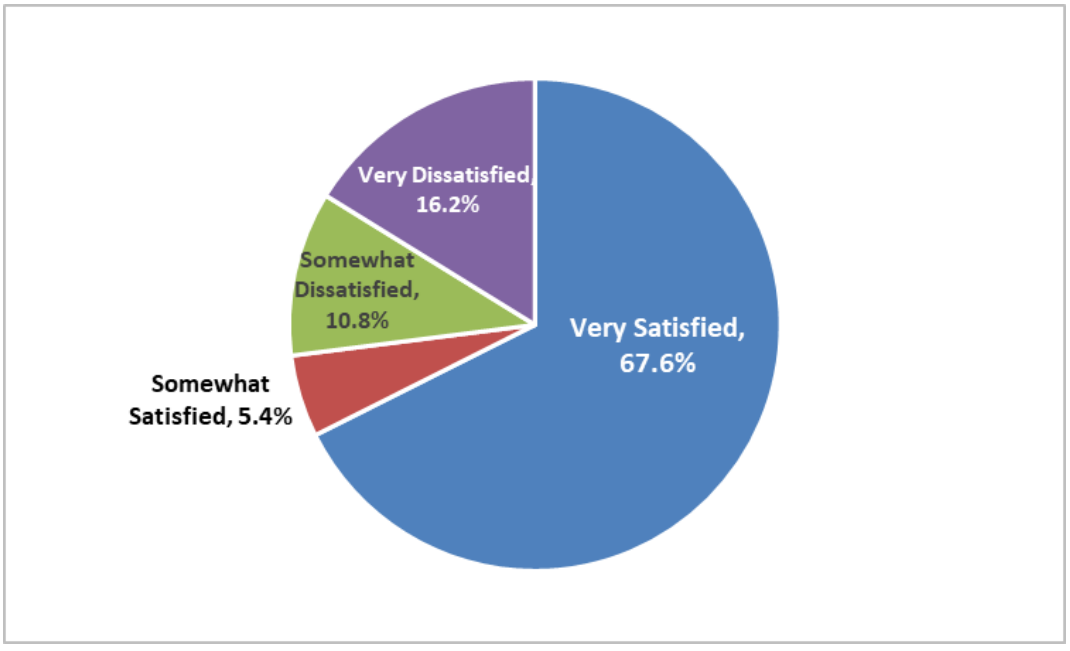
A majority of respondents who visited the emergency room in the past 12 months were "very" or "somewhat" satisfied with each of the aspects of care, but significant numbers were "somewhat" or "very" dissatisfied. In particular, 27 percent were dissatisfied in terms of being listened to and understood about managing their (or their child's) pain (Exhibit C-16).

Exhibit C-16 – Member Satisfaction with Aspects of Care (Emergency Room)

Aspect of Emergency Room Care	Very Satisfied	Somewhat Satisfied	Somewhat Dissatisfied	Very Dissatisfied	Not Sure
1. Seeing doctors who know how to care for patients with SCD	62.2%	10.8%	8.1%	16.2%	2.7%
2. Getting the right services and treatments, other than medication, for your (your child's) SCD	67.6%	13.5%	2.7%	16.2%	---
3. Being listened to and understood when you have concerns about managing your (your child's) SCD	67.6%	5.4%	5.4	21.6%	---
4. Getting the right medications to control your (your child's) pain	70.3%	5.4%	8.1%	16.2%	---

Overall satisfaction with care in the emergency room was consistent with ratings for individual aspects of care (Exhibit C-17). Over one-in-four respondents reported being "somewhat" or "very" dissatisfied with their care.

Exhibit C-17– Overall Satisfaction with Emergency Room Care



Members also were invited to describe their experiences with emergency room care. Several respondents described the challenges they confront when seeking treatment for their pain.

> *"Other than the OK City E.R., the emergency room doctors are arrogant and push people through. The doctors got offended when I tried to show them a letter from my son's specialists about his condition. They would not even read it telling me that they know what they are doing. They refused to listen to me or my son about his condition. They sent him home before he was ready, he told them he wasn't ready, and he ended up back in there after getting home."*

> *"My biggest complaint is with the E.R. doctors. Every time we are there the doctors talk down to us. I don't want to say it is a racial issue, but most Sickle Cell patients are African American and most of the doctors in the E.R. we see are not. They come across as unrelatable and detached…They need to have more compassion and understanding too. Patients with Sickle Cell, and their parents, have been through a lot for a lot of years. The doctors talk to us like it is the first time we have every heard of the disease.*

> *"I honestly do not think the doctors in the emergency room have any idea of the excruciating pain – pain the rest of us will never know – these kids are going through. Yet, the doctors act like we are in there seeking drugs for no reason."*

> *"I cannot get treatment for my Sickle Cell disease…I stopped even going to the E.R. because it is a waste of time. Sometimes I just curl up in my bed and cry from the pain."*

Town Hall

Town Hall attendees described similar experiences and expressed the same frustrations as survey respondents.

> *"I feel like it's always a fight, never a smooth process...ER visits are still kind of crazy. I'm still not comfortable and I kind of use it as my last resort. You know, I get doctors or nurses coming in, "What is sickle cell?" basically, or asking me basic questions they could've gotten off the internet. It puts me in an uncomfortable situation to be like my care is in your hands and you all are coming in asking me these simple questions. And, on top of that then (is the) pain crisis."*

"As a caregiver of a child with sickle cell, boy is it ever so difficult here…I have a 16-year-old that'd rather stay home than go to the ER… And, we really do worry about the difficulties she's going to have getting pain medications because when you're older seeking medication, they think you're drug seekers and will give you a harder time than ever to get pain meds."

It's always an experiment of what we could do from going to the ER, being left on the table because they think you're just pain drug seeking, to a nurse coming in and telling me to shut up because you're being too loud. That just happened recently which is why I don't like going to the ER because it's always that type of a factor. However, when you have do have a doctor and you actually have a relationship with that doctor, they tend to understand your needs are real. You have to kind of build that relationship with that doctor. And, you try and go to the hospital where your doctor is at. The doctor that I usually go to moved to another place so I'm stuck by myself with people that don't know what's going on, so you get those accusations and that makes it more difficult. Recently I spent about a week in the hospital. It was better because I went to the hospital where my doctor is so he was able to advocate for me. I try not to go to the ER by myself. I try to go with someone, like my mom, who can coherently speak. Trying to play chess with the doctors and nurses to let them know what's going on while in pain is cumbersome. Things are getting better because of building relationships over time with the doctors and nurses and with the information that is getting out there that this is real. So, it is getting better, but yes, the fight is real, it's strong."

Emergency Room Care – Summary of Findings

PHPG's review of the SoonerCare program's performance with respect to emergency room services found:

- Members with SCD are high utilizers of the emergency room because of the need for pain crisis treatment in a hospital setting.
- There are no statutory or Medicaid policy barriers preventing emergency room providers from treating patients in crisis aggressively, in terms of pain management.
- Emergency room providers at the Oklahoma centers-of-excellence rely on evidence-based protocols for treatment of patients in crisis and are familiar with the needs of members with SCD.

- Many providers in the State see patients in crisis infrequently and may not be equipped to treat pain promptly or aggressively enough, which can result in a lengthier and more severe crisis episode.
- Members with SCD express frustration and, in some cases, avoid seeking medically necessary care in the emergency room, out of fear of being labeled a drug seeker.

Evidence-based protocols for treatment of patients in crisis are readily available by emergency medical providers (see Appendix 6 for two sample protocols). These protocols emphasize the importance of early pain assessment and intervention.

The OHCA, in collaboration with centers-of-excellence and Supporters of Families with Sickle Cell Disease could undertake an educational campaign similar to the one discussed for PCMH providers in the previous section. The target of the campaign could include both emergency room physician groups and hospitals in counties where members with SCD reside.

The educational campaign need not be limited to emergency room physicians but also should include other staff (e.g., front desk personnel and triage nurses) who interact with patients. The information for these staff can be at a higher level, while imparting essential information, as demonstrated by the CDC flyer on the following page (Exhibit C-18).

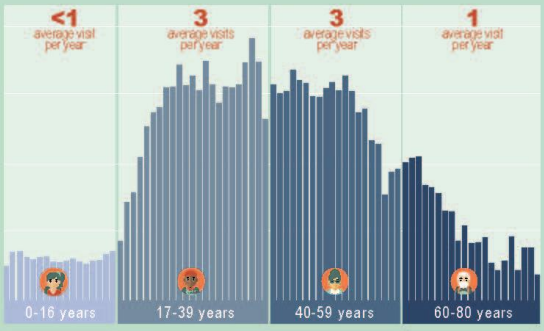
Exhibit C-18 – CDC Educational Materials

3 TIPS ABOUT SICKLE CELL DISEASE

EVERY EMERGENCY PROVIDER NEEDS TO KNOW

Children and adults with sickle cell disease (SCD) often require care in the emergency department (ED) of hospitals and clinics for health issues related to SCD. The ED may be a patient's only option for health care when symptoms, such as pain crises, cannot be managed at home or when a patient does not have access to a healthcare provider who specializes in treating SCD. The Sickle Cell Data Collection (SCDC) program found that in California, people with SCD seek care in the ED an average of three times a year from their late teens to their late 50s.

Emergency Department (ED) Visits Among People with Sickle Cell in California, 2005-2014



Tips for ED Health Providers

- Take complaints of pain from patients with SCD seriously and treat promptly with appropriate fluids and pain medication.
- Work with the SCD team at your hospital or clinic to develop individualized care plans for patients with SCD, especially those with frequent ED use. When possible, make these plans available in the electronic medical record.
- Refer to the National Heart, Lung, and Blood Institute guidelines for the management of SCD: *www.nhlbi.nih.gov/health-pro/guidelines/sickle-cell-disease-guidelines*

Primary Health Complaint: Extreme Pain

Pain crises, which can be excruciating, are the most common reason for ED visits among patients with SCD. Patients may not always appear to be in pain because they have often developed a high pain tolerance due to a lifetime of chronic pain.

Patients with SCD require prompt pain treatment. The medical evaluation of patients includes determining the cause of pain and assessing recent medication use. For mild or moderate pain, begin treatment with nonsteroidal anti-inflammatory drugs. For severe pain, treatment with opioids may be needed. If the patient is already on opioid therapy, calculate opioid dose based on current opioid dose. Reassess pain and provide additional opioid administration, if necessary, for continued severe pain. For greater effectiveness, medication can be combined with nonpharmacologic approaches, such as heat application and distraction.

PROTECTING PEOPLE

CDC's National Center on Birth Defects and Developmental Disabilities is committed to protecting people and preventing complications of blood disorders. Learn more about CDC's work to help people with SCD here: **www.cdc.gov/ncbddd/sicklecell**

U.S. Department of Health and Human Services
Centers for Disease Control and Prevention

SCDC
Sickle Cell Data Collection

A best practice described in literature and recommended by Supporters of Families with Sickle Cell Disease is creation of a pain management action plan. The plan is a written description from the patient's Hematologist that outlines his or her condition, needs and recommended course-of-care when in crisis[18]. Patients with an action plan have a better (though not certain) chance of receiving timely care when they arrive in an unfamiliar emergency room. The action plans also could be uploaded to the State HIE for ready access from any hospital.

The OHCA, SoonerSelect MCEs and advocacy community should collaborate on outreach to members with SCD and Hematology community to facilitate creation of plans. Emergency room providers also can be educated on their efficacy and importance.

[18] The majority of survey respondents reported having a pain management plan but PHPG did not frame the question specifically to ask about an action plan with Hematologist-recommended steps in the event of a crisis for review by an emergency room physician.

4. Supports for Members with SCD to Navigate the System

PHPG evaluated available supports for members with SCD to navigate the health care system through:

- Literature review of best practices
- Review of existing SoonerCare care management initiatives
- Review of SoonerSelect care management requirements

Documented Best Practices

Individuals with a complex/chronic disease such as SCD often require care from multiple medical specialties, as well as behavioral health services to cope with what is a life-long condition. Navigating the health care system without support can lead to fragmented care or gaps in care, as well as patient discouragement.

One recognized best practice for managing complex care needs is through establishment of a member-centered interdisciplinary care team[19]. The team typically includes representatives from all specialties relevant to the individual's health needs, both interventionist and palliative, as well as a designated care manager (usually a nurse or social worker) to coordinate the team's activities. As suggested by its name, the team places the member at its center, and s/he retains autonomy for choosing the preferred course of care.

Medicaid beneficiaries often face additional, non-clinical hurdles to accessing care. These factors, known as "social determinants of health" (SDOH) can include housing insecurity, food insecurity, difficulty making utility payments and lack of reliable transportation, among others. A person with significant SDOH needs may, by necessity, regard their health care needs, particularly preventive health care, as a lesser priority.

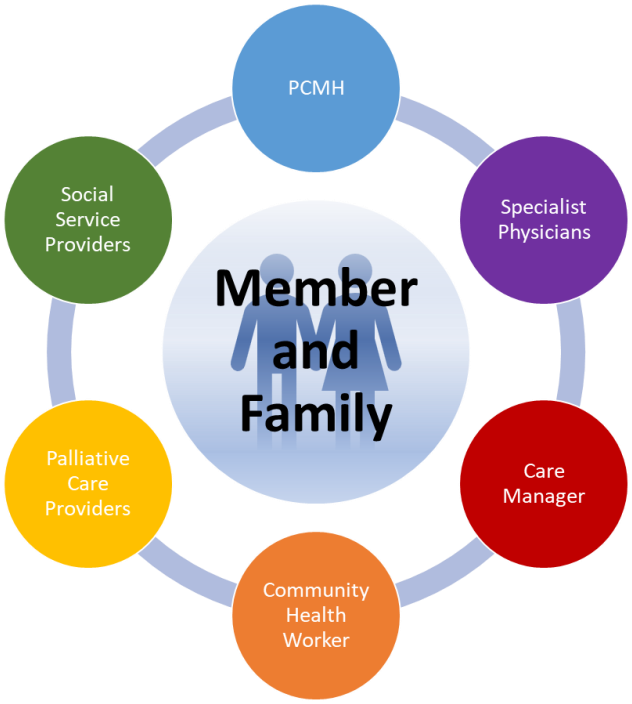
Individuals enrolled with an interdisciplinary care team typically receive a comprehensive assessment, followed by creation of a care plan that addresses both clinical and non-clinical (social) needs. Preventive and therapeutic services are addressed, with preventive services for members with SCD encompassing condition-specific interventions, such as

[19] For a discussion of the benefits of the interdisciplinary team model, see: Implementation of an Interdisciplinary, Team-Based Complex Care Support Health Care Model at an Academic Medical Center: Impact on Health Care Utilization and Quality of Life, Implementation of an Interdisciplinary, Team-Based Complex Care Support Health Care Model at an Academic Medical Center: Impact on Health Care Utilization and Quality of Life | PLOS ONE

hydration therapy or blood transfusions (as applicable). SDOH needs may be managed by a Community Health Worker trained for this task[20].

The interdisciplinary care team can facilitate the transition from pediatric to adult care by assessing a member's readiness to transition, planning for the transition and facilitating the transfer. Peer supports also can be made available, if desired by the member. The composition of the team can evolve concurrently, with adult providers replacing their pediatric counterparts as appropriate (Exhibit C-19).

Exhibit C-19– Interdisciplinary Care Team Model



The surveyed providers strongly endorsed the value of the activities performed by an interdisciplinary care team. Nearly all of the respondents said it would be "very helpful" for persons with SCD to receive education and counseling, as well community-based supports and assistance with transition planning (for adolescents).

There also is a growing body of research on the value of using mobile applications (smart phone technology) to support monitoring of patients with chronic conditions, including SCD. A 2018-2019 Agency for Healthcare Research and Quality-funded randomized study conducted at Duke University on 59 patients with SCD being discharged from the hospital found that:

[20] For a description of the Community Health Worker function within an interdisciplinary care team, see: Addressing Social Determinants of Health through Community Health Workers: A Call to Action, HHC-CHW-SDOH-Policy-Briefi-1.30.18.pdf (cthealth.org)

Patients using SMART had significantly less acute care utilization and were more likely to return for follow up visits. The use of a simple technology solution such as a mobile app to record symptoms, allowed symptoms such as pain to be reviewed remotely. Daily review of pain scores remotely provided the medical team with the ability to text specific patients believed to be at risk due to increasing pain. To aid in follow up, SMART also included the ability to have a reminder for an appointment 'pop-up'. Technology reminders also led to patients being more likely to return as scheduled for their appointment as compared to standard-of-care (control group)[21].

A mobile app could have particular appeal to younger members with SCD. Individuals who both are enrolled with an interdisciplinary care team and equipped with a mobile app would be well-supported when navigating the health care system.

OHCA Care Management – Current and SoonerSelect

There are multiple pathways through which SoonerCare members with SCD can receive care management today. Members who are seen at Jimmy Everest have access to an interdisciplinary care team in accordance with best practices. In addition to addressing current needs, the team assists members to prepare for the transition from pediatric to adult care and coverage.

A small number of members with significant care needs also are enrolled in the SoonerCare HMP or receive care management through a SoonerCare Health Access Network. And members not otherwise being care managed are identified for enrollment in the OHCA's Chronic Care Unit if they meet pre-established utilization/expenditure thresholds. The CCU offers important assistance, including with transition from pediatric to adult coverage, if applicable.

Members eligible for long term care also receive active care management as a component of their services. (See page 18 for an illustration of current and planned SoonerCare care management options.)

PHPG asked survey respondents whether they had been assigned a care or case manager. Seventeen respondents (27.4 percent) answered yes, while 38 (61.3 percent) answered no. (Seven respondents (11.3 percent) were unsure.)

SoonerSelect will expand access to interdisciplinary care management. MCEs will be required to offer an initial health screening to all new enrollees, and to perform a comprehensive clinical and SDOH assessment on those identified as having special needs, a category that would include members with SCD. The assessment will be used to develop

[21] SMART Mobile Application Technology Utilization in the Treatment of Sickle Cell Disease Post Day Hospital Discharge - Full Text View - ClinicalTrials.gov

a comprehensive, interdisciplinary care plan, to be overseen by a designated care manager[22].

Access to Supports – Summary of Findings

PHPG's review of the SoonerCare program's performance with regard to member supports found:

- Members with complex/chronic conditions can benefit from access to an interdisciplinary care team that addresses both clinical and social needs.
- Mobile app technology can offer an additional tool for monitoring a member's health status and adherence to preventive care guidelines.
- The SoonerCare program currently offers care management to some members with SCD, although the majority do not have an assigned care manager or interdisciplinary care team.
- All members with special needs who are enrolled in SoonerSelect will have access to a care manager and care team.

The majority of SoonerCare Choice members with SCD will transition to SoonerSelect in April 2024. The residual population will include non-Medicare eligible ABD beneficiaries, some of whom already receive care management through the CCU, HMP or a HAN.

The OHCA has the opportunity to ensure all members with SCD have access to care management (ideally through an interdisciplinary care team), including assistance with transition to adult coverage, by targeting ABD beneficiaries not currently assigned a care manager. The OHCA should collaborate with the HMP vendor and HANs to contact all non-care managed ABD members for the purpose of performing an assessment and developing a care plan, as appropriate. Special emphasis should be placed on reaching and engaging members with very complex needs (e.g., members with other chronic conditions related to their SCD and members with frequent inpatient hospital admissions) who do not yet have a care manager.

The OHCA also should ensure that current and future care management systems emphasize the importance to members of having an action plan that addresses pain management and other priorities. The team, through the member's care manager, should be available to consult on an urgent basis with emergency room physicians when a member in crisis presents for care.

The OHCA should explore the use of mobile app technology as an additional care management tool. Such an app could be operated directly by the OHCA or through the SoonerSelect MCEs, the SoonerCare HMP vendor and SoonerCare HANs.

[22] SoonerSelect Medical RFP, Section 1.9.3, Care Management and Population Health

D. SUMMARY OF RECOMMENDATIONS

SoonerCare members with SCD face challenges in managing a complex, and in many instances, extraordinarily painful condition. Parents of children with SCD have the added burden of advocating on behalf of someone who may be unable to articulate fully his or her needs.

The SoonerCare program has acted to assist members with SCD, both through its existing care management systems and its contract with Supporters of Families with SCD. The OHCA's SoonerSelect program will expand access to care management for members with SCD who are enrolled into an MCE.

The two centers-of-excellence in the State provide expert care to the members they serve. Together, they account for a significant percentage of all SoonerCare members with SCD in Oklahoma.

However, significant opportunities for improvement exist. PCMH and emergency room providers unfamiliar with SCD can be unaware of evidence-based guidelines for care, particularly for patients experiencing a pain crisis. Members without access to a care manager also may have difficulty navigating what is a complex health care system.

Exhibit D-1 below summarizes findings and recommendations from Report Section C. All of the recommendations are actionable in the next 12 to 18 months. The second annual report, due to the Legislature in January 2024, will provide an update on the status of the recommendations.

Exhibit D-1 – Findings and Recommendations

Findings (Areas for Improvement)	Recommendations
Access to Care	
1. PCMH providers are not uniformly knowledgeable about Sickle Cell Disease.	1. The OHCA, in conjunction with SoonerSelect MCEs, centers-of-excellence and Supporters of Families with SCD, should conduct a coordinated educational campaign targeting PCMH providers in counties with SCD members. PCMH providers could be offered continuing education credits for participating.
2. Specialist providers, including at centers-of-excellence, must channel referrals through PCMH providers.	2. The OHCA and SoonerSelect MCEs should have a process for allowing qualified specialists to make referrals, while keeping PCMH providers informed, either directly or through the HIE.

Findings (Areas for Improvement)	Recommendations
Emergency Room Provider Training and Resources	
3. Many ER providers see patients in crisis infrequently and may not be equipped to treat pain promptly or aggressively.	3. The OHCA, in collaboration with centers-of-excellence and Supporters of Families with Sickle Cell Disease, should undertake an educational campaign to increase knowledge of evidence-based protocols for treatment.
4. Only a portion of members with SCD today have a pain management action plan.	4. The OHCA, SoonerSelect MCEs and advocacy community should collaborate on outreach to members with SCD and Hematology community to facilitate creation of plans. Emergency room providers also can be educated on their efficacy and importance.
Supports of Members with SCD to Navigate the Health Care System	
5. Only a portion of members with SCD today are assessed to identify the potential need for care management.	5. The OHCA should collaborate with the HMP vendor and HANs to contact all non-care managed ABD members for the purpose of performing an assessment and developing a member-centered care plan, as appropriate. Members with very complex conditions (e.g., members with other chronic conditions due to SCD (e.g., kidney disease) or members with frequent inpatient hospital admissions) who are not yet engaged should be given top priority. (All SoonerSelect members will be assessed at time of enrollment.)
6. Only a portion of members with SCD today have a comprehensive action or care plan that addresses both clinical and social service needs.	6. The OHCA also should ensure that current and future care management systems emphasize the importance to members of having a comprehensive care/action plan that addresses the member's complete care needs, both current and future for members transitioning from pediatric to adult coverage.
7. Members enrolled in care management should have access to an interdisciplinary care team, as appropriate.	7. The OHCA should strive to make available interdisciplinary care management, where appropriate, by coordinating with the Jimmy Everest Center, SoonerCare HMP and SoonerCare HANs on behalf of ABD beneficiaries. (All SoonerSelect enrollees will have access to interdisciplinary care teams.)

Findings (Areas for Improvement)	Recommendations
8. Interdisciplinary care teams are well-suited to facilitating a member's transition from pediatric to adult care.	8. The OHCA should ensure that SoonerSelect and other care management systems target adolescents and assist in the transition to adult coverage and care.
9. Mobile app technology can offer an additional means of supporting members.	9. The OHCA should explore use of a mobile app, either directly or through its contractors.

E. APPENDICES

Report appendices are presented, starting on the following page. The appendices include:

Appendix	Content
Appendix 1	Selected References
Appendix 2	Virtual Town Hall meeting invitation
Appendix 3	PHPG member survey instrument
Appendix 4	OHCA out-of-State services prior authorization form
Appendix 5	Hospital admitting diagnosis data
Appendix 6	Sample emergency room protocols

1. Selected References

Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action 2020, The National Academies of Sciences, Engineering and Medicine, Addressing Sickle Cell Disease A Strategic Plan and Blueprint for Action | National Academies

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Implementation of an Emergency Department Screening and Care Management Referral Process for Patients with Sickle Cell Disease, Professional Case Management (September/October 2019), Implementation of an Emergency Department Screening and Care Management Referral Process for Patients With Sickle Cell Disease - PubMed (nih.gov)

Improving Sickle Cell Transitions of Care through Health Information Technology: Recommendations for Tool Development, American Journal of Preventive Medicine (July 2016), Improving Sickle Cell Transitions of Care Through Health Information Technology - PubMed (nih.gov)

A Program of Transition to Adult Care for Sickle Cell Disease, American Society of Hematology (December 2019), A program of transition to adult care for sickle cell disease - PubMed (nih.gov)

Emergency department management of acute pain episodes in sickle cell disease, Academic Emergency Medicine (March 2007), Emergency department management of acute pain episodes in sickle cell disease - PubMed (nih.gov)

Adult emergency department patients with sickle cell pain crisis: a learning collaborative model to improve analgesic management, Academic Emergency Medicine (April 2010), Adult emergency department patients with sickle cell pain crisis: a learning collaborative model to improve analgesic management - PubMed (nih.gov)

(See also the publications cited directly in body of the report.)

2. *Virtual Town Hall Meeting Invitation*

3. PHPG Member Survey Instrument

SoonerCare Member Survey
Members/Families with SCD

Hello, my name is _____ and I am calling on behalf of the SoonerCare program. May I please speak to {RESPONDENT NAME}?

INTRO **We are conducting a short survey to find out about where SoonerCare members get their health care and their experiences with doctors and nurses. The purpose of the survey is to learn about how we can make the program better. The survey is voluntary and if you decide not to participate it will not affect your benefits. Anything you tell us will be kept confidential. The information will not be shared with your doctor or nurse and will not affect any treatment you may be receiving. The survey takes about 10 minutes.**

[ANSWER ANY QUESTIONS AND PROCEED TO QUESTION 1]

Parent/Guardian Screening Question: Are you the parent or guardian of [NAME]? [IF YES, PROCEED TO QUESTION 1. IF NO, ASK TO SPEAK TO PARENT/GUARDIAN. IF UNABLE TO REACH, END CALL]

Screening for Medicaid

1. The SoonerCare program is a health insurance program offered by the state. Are you currently enrolled in SoonerCare?[23] [IF MINOR → Is [NAME] currently enrolled in SoonerCare?]
    a. Yes
    b. No → [ASK IF ENROLLED IN MEDICAID. IF NO, END CALL]
    c. Don't Know/Not Sure → [ASK IF ENROLLED IN MEDICAID. IF NO, END CALL]

2. We are interested in talking today to people (parents of children) who are being treated for Sickle Cell Disease, including Sickle Cell Anemia. Are you (Is your child) being treated for this condition? [IF MULTIPLE CHILDREN, CLARIFY THAT SURVEY QUESTIONS ARE TO BE ANSWERED WITH RESPECT TO THE CHILD NAMED AT THE OUTSET OF THE CALL.] [IF YES, SCA OR OTHER]
    a. Yes, Sickle Cell Anemia
    b. Yes, other Sickle Cell Disorder
    c. No → [END CALL]

[23] Select questions include a "don't know/not sure" or similar option which is unprompted by the surveyor; this response is listed on the instrument to allow surveyors to document such a response. Questions are reworded for parents/guardians completing the survey on behalf of program participants.

Regular Care

3. First, I'm going to ask a few questions about where you (your child) get your regular care. Do you (does your child) have a regular doctor or nurse practitioner, someone you go to for check-ups, when you need advice about a health problem or get sick or hurt? [IF RESPONDENT HAS MORE THAN ONE PROVIDER, CODE AS YES].
    a. Yes
    b. No → [GO TO Q5]
    c. Don't Know/Not Sure → [GO TO Q5]

4. Is your regular doctor or nurse practitioner a primary care provider, like a family physician, general internist or pediatrician? [IF YES, RECORD TYPE] Or is she or he a specialist? [IF SPECIALIST] What is your regular provider's specialty?
    a. Family physician/nurse practitioner → [GO TO Q6]
    b. General internist → [GO TO Q6]
    c. Pediatrician → [GO TO Q6]
    d. Specialist – Hematology → [GO TO Q7]
    e. Specialist – Other [SPECIFY]
    f. Don't Know/Not Sure

5. Where do you usually go to get health care (health care for your child)? → [RECORD AND GO TO Q7]
    a. Emergency Room
    b. Urgent Care Clinic
    c. No usual place
    d. Have never tried to get care
    e. Don't Know/Not Sure

6. Do you have a doctor, other than your (your child's) regular provider, who helps you manage your (your child's) Sickle Cell Disease? [IF YES] What is this doctor's specialty?
    a. Yes – Hematology
    b. Yes – Other [SPECIFY]
    c. Yes – Don't Know/Not Sure of specialty
    d. No
    e. Don't Know/Not Sure

7. Some SoonerCare members with complicated medical conditions are assigned care or case managers to provide extra help between doctor's visits. These are nurses who might work for the Oklahoma Health Care Authority or be part of your doctor's care team. Do you (does your child) have a care manager or case manager?
    a. Yes
    b. No
    c. Don't Know/Not Sure

8. Next, I'm going to ask you a few questions about some types of care you (your child) might receive to treat or manage your (his/her) Sickle Cell Disease. For each question, please tell me if you (your child):

	Yes	No	DK/Not Sure	Not Applicable
a. [IF MEMBER IS YOUNGER THAN 5 YEARS] Receives daily penicillin				
b. [IF MEMBER IS OLDER THAN 9 MONTHS] Receives Hydroxyurea medication				
c. [IF MEMBER HAS SCA & IS AGE 2 TO 16] Had a head X-ray called a Transcranial Doppler at least once in the past twelve months				
d. [IF MEMBER IS 3 OR OLDER] Had blood pressure checked at least once in the past twelve months				
e. [IF MEMBER IS 10 OR OLDER] Had retinas (eyes) examined at least once in the past twelve months				
f. [IF MEMBER IS 12 MONTHS OR OLDER] Had kidneys checked at least once in the past twelve months				
g. Had lungs checked at least once in the past twelve months				
h. Had heart checked at least once in the past twelve months				
i. Received a pneumococcal vaccine (shot to prevent pneumonia) in the past twelve months				
j. Received one or more blood transfusions in the past twelve months				
k. Currently receive chelation therapy for iron overload				
l. Have a treatment plan for pain control				

9. Next, I'm going to ask you a few questions about how satisfied you are with the care you (your child) receive(s) for treatment of your (his/her) Sickle Cell Disease from your regular doctors and other members of your care team. This does not include doctors you might have seen in an emergency room. For each question, please tell me if you are Very Satisfied, Somewhat Satisfied, Somewhat Dissatisfied or Very Dissatisfied with your care.

	Very Satisfied	Somewhat Satisfied	Somewhat Dissatisfied	Very Dissatisfied	DK/Not Sure
a. Finding doctors who know how to care for patients with Sickle Cell Disease					
b. Being able to schedule doctor's appointments when you need them for treatment of your (your child's) Sickle Cell Disease					
c. Getting the right services and treatments, other than medication, for your (your child's) Sickle Cell Disease					
d. Getting the right medications for your (your child's) Sickle Cell Disease					
e. Getting extra support and help if you need it to manage your Sickle Cell Disease					
f. Being listened to and understood when you have concerns about your (your child's) Sickle Cell care in general, other than pain management					
g. Being listened to and understood when you have concerns about managing your (your child's) Sickle Cell pain					

10. Overall, how satisfied are you with the care you (your child) receive from your regular doctors and other members of your care team for treatment of your (your child's) Sickle Cell Disease? Would you say you are Very Satisfied, Somewhat Satisfied, Somewhat Dissatisfied or Very Dissatisfied?
    a. Very satisfied
    b. Somewhat satisfied
    c. Somewhat dissatisfied
    d. Very dissatisfied
    e. Don't Know/Not Sure

11. Why did you choose that answer? [RECORD]

12. [CAHPS QUESTION – FOR COMPARISON TO OTHER CARE MANAGED POPULATIONS] And using any number from 0 to 10, where 0 is the worst health care possible and 10 is the best health care possible, what number would you use to rate all your health care in the last six months? [RECORD]

Emergency Room Care

13. Have you (has your child) been treated in an emergency room in the past 12 months for your (your child's) Sickle Cell Disease? [IF YES] About how many times in the past 12 months?
    a. No visits → [GO TO Q17]
    b. Yes [RECORD NUMBER]
    c. Don't Know/Not Sure → [GO TO Q17]

14. Next, I'm going to ask you a few questions about how satisfied you are with the care you (your child) receive(s) for treatment of your (his/her) Sickle Cell Disease from doctors in the emergency room. If you had more than one visit in the past 12 months, think just about your most recent visit. For each question, please tell me if you are Very Satisfied, Somewhat Satisfied, Somewhat Dissatisfied or Very Dissatisfied with your care.

	Very Satisfied	Somewhat Satisfied	Somewhat Dissatisfied	Very Dissatisfied	DK/Not Sure
a. Seeing doctors who know how to care for patients with Sickle Cell Disease					
b. Getting the right services and treatments, other than medication, for your (your child's) Sickle Cell Disease					
c. Being listened to and understood when you have concerns about managing your (your child's) Sickle Cell pain					
d. Getting the right medications to control your (your child's) pain					

15. Overall, how satisfied are you with the care you (your child) received in the emergency room on your most recent visit for treatment of your (his/her) Sickle Cell Disease? Would you say you are Very Satisfied, Somewhat Satisfied, Somewhat Dissatisfied or Very Dissatisfied?
    a. Very satisfied
    b. Somewhat satisfied
    c. Somewhat dissatisfied
    d. Very dissatisfied
    e. Don't Know/Not Sure

16. Why did you choose that answer? [RECORD]

Out-of-State Care

17. Have you tried to see providers outside Oklahoma who specialize in treating Sickle Cell Disease since you (your child) became a SoonerCare member?
    a. Yes
    b. No → [GO TO Q23]
    c. Don't Know/Not Sure → [GO TO Q23]

18. What provider or providers did you try to see? [RECORD]

19. How did you learn about this provider (these providers)? [RECORD ALL ANSWERS]
    a. From one of my regular doctors
    b. From an emergency room doctor
    c. From my own research/internet
    d. From a Sickle Cell Disease support group [RECORD NAME]
    e. Other [RECORD]
    f. Don't Know/Not Sure

20. Why did you want to see a provider outside of Oklahoma? [IF MULTIPLE REASONS] What was the most important reason? [RECORD MOST IMPORTANT REASON]
    a. Not satisfied with Oklahoma provider access
    b. Not satisfied with Oklahoma provider knowledge of Sickle Cell Disease
    c. I wanted a service or treatment not available in Oklahoma [RECORD SERVICE OR TREATMENT]
    d. It was more convenient to my place of residence
    e. Other [RECORD]
    f. Don't Know/Not Sure

21. Did you or your provider ask the Oklahoma Health Care Authority to approve and pay for you to receive care out-of-state? [IF YES] Was some or all of it approved?
    a. Yes – all approved → [GO TO Q23]
    b. Yes – some approved and some denied
    c. Yes – all not approved/denied
    d. No – did not ask for approval → [GO TO Q23]
    e. Still awaiting determination → [GO TO Q23]
    f. Don't Know/Not Sure → [GO TO Q23]

22. [IF PARTIAL] For the services that were denied, did you see the out-of-state provider(s) and arrange payment on your own? [IF ALL DENIED] Did you see the out-of-state provider(s) and arrange payment on your own?
    a. Yes
    b. No
    c. Don't Know/Not Sure

Transition-of-Care

[IF RECORD SHOWS SURVEY SUBJECT IS 14 TO 19 YEARS OF AGE, ASK TOC QUESTIONS 22 – 24. IF RESPONDENT IS 14 TO 17, ALSO ASK QUESTION 25]

I have just a few more questions.

23. Would you say that your child receives the majority of his/her care in a pediatric setting or an adult care setting?
    a. Pediatric
    b. Adult
    c. Mixed
    d. No regular setting
    e. Don't Know/Not Sure

24. [IF ADULT SETTING – DID ANYONE] Has anyone on your child's care team or at the state help(ed) you to prepare for your child's transition from receiving care in a pediatric setting to an adult setting? [IF YES] Who has talked to you? [RECORD ALL]
    a. Yes – Doctor/care team
    b. Yes – State
    c. Yes - Both
    d. No → [GO TO Q26 IF AGE 14-17; ELSE GO TO 27]
    e. N/A – child receives care in an adult setting/from adult care provider(s) → [GO TO Q26 IF AGE 14-17; ELSE GO TO 27]
    f. Don't Know/Not Sure → [GO TO Q26 IF AGE 14-17; ELSE GO TO 27]

25. How did they help you prepare? [RECORD]

26. [ASK IF AGE 14 – 17] Do you feel confident you know how your child's SoonerCare coverage might change when she/he turns 18?
    a. Yes
    b. No
    c. Don't Know (if confident)/Not Sure

Suggestions

27. Do you have any suggestions for how the SoonerCare program could do a better job helping you manage your (your child's) Sickle Cell Disease? [RECORD ALL]

28. [IF TWO OR MORE SUGGESTIONS] Thinking about the suggestions you just made, which one suggestion would you say is the most important? [RECORD]

Those are all my questions. Thank you for your time.

4. OHCA Out-of-State Services PA Form and Notice

SoonerCare
Oklahoma Health Care Authority

HCA-65

Out-of-State Prior Authorization Request

Member Name _____ ☐ Male ☐ Female RID _____ DOB _____

Member Address _____

Parent/Caregiver _____ Contact Number _____

Referring PCP or Specialist _____ NPI or Provider ID # _____

Contact for referring Provider _____ Contact Number _____

Active Diagnosis/Diagnoses Related to Request _____

Services Being Requested _____

Are these services emergent or urgent based on clinical conditions? If yes, why?

Date of Service _____ ☐ Outpatient ☐ Inpatient Length of Stay (Inpatient) _____

Is transportation needed? If so, what type? _____

Accepting Provider/Facility _____ Provider ID# or NPI _____

Full Address _____ Phone _____

FOR OHCA USE ONLY Non-Contracted _____ Willing to Contract _____

Accepting Provider/Facility _____ Provider ID# or NPI _____

Full Address _____ Contact Person _____ Phone _____

FOR OHCA USE ONLY Non-Contracted _____ Willing to Contract _____

Please attach the following:

- Documentation to establish the medical necessity of services requested, such as medical records
- Letter of medical necessity or other thorough summary document that includes:
  - Summary of the member's condition and history of treatment related to request
  - History of other providers who have evaluated, treated or consulted member related to request
  - Recommended treatment or further diagnostic needed
  - Why medical care cannot be completed in Oklahoma or the next closest location

Please fax the completed request to OHCA Population Care Management at (405) 530-3217.

Date Received by OHCA _____

4345 N. Lincoln Blvd., OKC | 800-522-0114 | okhca.org |

OKLAHOMA
Health Care Authority

SoonerCare Out-of-State
SERVICES RULE CHANGES

Beginning Sept. 1, 2019 the Oklahoma Health Care Authority (OHCA) will enact changes to the agency's out-of-state (OOS) services policies. These changes will continue to ensure members have access to quality care while controlling program costs. These changes will not impact routine medical care for SoonerCare members.

In 2019 the Oklahoma legislature passed HB 2341, which limited SoonerCare members' services to in-state providers when possible. These changes to OOS services will allow OHCA to maintain compliance with federal and state regulations.

These revisions clearly define coverage and reimbursement for services rendered by providers that are physically located outside of Oklahoma. The policy also outlines provider participation requirements and documentation requirements for OOS service requests.

WHAT **SOONERCARE MEMBERS** NEED TO KNOW

- Members living near the Oklahoma state border, who regularly see a SoonerCare-contracted provider across the border, should see no changes, as long as the provider's office is within 50 miles of the Oklahoma border. In the case of a provider's office being more than 50 miles away from the Oklahoma border, an out of state prior-authorization will be required.
- Medical care needed due to an accident or medical emergency while a member is travelling in another state is still eligible for compensation once medical necessity is determined.
- Single-case agreements and contracts will not be allowed under the rule changes. SoonerCare members currently receiving OOS services through single-case agreements will be transitioned to regularly-contracted SoonerCare providers that OHCA medical staff have determined can provide the same level of care at OHCA's regularly contracted rates.
- Self-referrals will no longer be permitted and members will be responsible for incurred medical costs if they do not receive the proper prior authorization for OOS services. Members who think they need out-of-state services should discuss the apparent need with their primary care provider.

WHAT **SOONERCARE PROVIDERS** NEED TO KNOW

- Except for behavioral health emergencies and true medical emergencies, all required prior authorization documentation must be received by OHCA 10 days in advance of the day the OOS services are to be rendered.
- Requests for care will not be evaluated until all required documents are completed and submitted to OHCA. See below for a list of required prior authorization documents.
- Referring providers should submit OHCA Form HCA-65 Out of State Prior Authorization Request with required documentation by fax to OHCA Population Care Management at 405-530-3217. This form can be found at http://www.okhca.org//oosproviders.

OKLAHOMA
Health Care Authority

WHAT **SOONERCARE PROVIDERS** NEED TO KNOW

- Members may not be sent to non-contracted providers, facilities or doctors. While the referring provider may suggest a destination for the member's treatment, the ultimate decision on destination will be made by the OHCA Chief Medical Officer or his/her designate and will be based on treatment consistent with recognized standards of care, cost effectiveness and contract status of providers.
- Telephone requests for OOS services will only be approved in true emergencies and must be followed promptly with the submission of all required documentation.
- For emergent telephone requests, please call Population Care Management at 877-252-6002.

DOCUMENTATION REQUIRED WITH PRIOR AUTHORIZATION APPLICATION

- Documents determining medical necessity for the procedure, such as history of present illness, past medical and surgical history, physical exam, lab and imaging reports, progress notes and other relevant documents.
- Letter of Medical Necessity (or other summary document) from the provider that contains:
  - *Member's condition and diagnosis, including a clear summary of treatment to date that is supported by the medical records.*
  - *Names of physicians and/for facilities in Oklahoma that the member has previously been referred to for diagnosis and/or treatment.*
  - *Physicians consulted by attending physician related to the diagnosis and/or availability of recommended treatment in Oklahoma.*
  - *Recommended treatment or further diagnostic work.*
  - *Reasons why care cannot be provided in Oklahoma.*

*Created September 3, 2019

ADDRESS
4345 N. Lincoln Blvd.
Oklahoma City, OK 73105

WEBSITES
okhca.org
mysoonercare.org

PHONE
Admin: 405-522-7300
Helpline: 800-987-7767

5. Hospital Admitting Diagnosis Data

Diagnosis Category	Admissions	Percent of Total
Sickle Cell Anemia	1,145	60.4%
Other Sickle Cell Disease	107	5.6%
Sepsis	75	4.0%
Other Diagnoses	56	3.0%
Pregnancy and Childbirth	51	2.7%
Newborn	48	2.5%
Diseases of the Digestive System	44	2.3%
Behavioral Health	42	2.2%
Diseases of the Musculoskeletal System	35	1.8%
Pneumonia	32	1.7%
Thalassemia	25	1.3%
Hemochromatosis	22	1.2%
Other Diseases of the Respiratory System	20	1.1%
COVID	19	1.0%
Infectious and Parasitic Diseases	18	0.9%
Other Diseases of the Genitourinary System	16	0.8%
Other Diseases of the Circulatory System	15	0.8%
Fever	14	0.7%
Diseases of the Nervous System	14	0.7%
Kidney Failure	14	0.7%
Bronchitis	12	0.6%
Hypertension	11	0.6%
Embolism	9	0.5%
Influenza	9	0.5%
Other Disorders of Fluid, Electrolyte and Acid-Base Balance	8	0.4%
Anemia	7	0.4%
Diabetes	7	0.4%
Diseases of the Skin	6	0.3%
Neoplasm	5	0.3%
Pharyngitis	5	0.3%
Volume Depletion	5	0.3%
Total	**1,896**	**100.0%**

6. Sample Emergency Room Protocols

New York City Order Set for Patients with Sickle Cell Disease

Appendix. Order Set for Patients with Sickle Cell Disease (SCD) Presenting to the Emergency Department (ED) with Pain in New York City

The following is a suggested order set for use in the ED. Except as noted, the listed elements are those which a panel of clinicians agreed would be very likely to improve outcomes. This order set is not intended to be implemented without local discussion. Consider each item carefully in the context of your ED.

A. TRIAGE

- ☐ Identify SCD patient and initiate SCD protocol
- ☐ Assess vitals, including O_2 sat
- ☐ Assess pain using VAS or verbal scale (1-10)[i]
- ☐ Confirm allergies to medications (opiates, NSAIDs, antibiotics, etc.)
- ☐ Assign ESI Level 2
- ☐ Begin implementation of rapid protocol (initiate analgesic therapy <30 minutes after triage)

B. INITIAL MEDICAL ENCOUNTER

- ☐ Review vitals (including O_2 sat)
- ☐ Assess pain using VAS or verbal scale (1-10)
- ☐ Note treatment prior to coming to ED or in triage (opioids, NSAIDs)
- ☐ Note baseline hemoglobin†
- ☐ Note date of and reaction to last transfusion†

Assess if patient has a documented SCD treatment plan:

- ☐ If yes, review with patient and integrate with items in sections **E [INITIAL PAIN MANAGEMENT]** and **F, G, H [PAIN REASSESSMENTS]**
- ☐ If no, attempt to find analgesic history during previous ED visits in medical record

- ☐ Confirm usual analgesic type and dose with patient

C. DRAW LABS

Draw labs as appropriate:

- ☐ CBC with differential
- ☐ Reticulocyte count
- ☐ Electrolytes (CHEM-7)
- ☐ ALT and AST†
- ☐ LDH
- ☐ Bilirubin
- ☐ Type and screen (if no active type and screen)
- ☐ Hemoglobin fractionation/electrophoresis†
- ☐ Iron studies (Fe, TIBC, Ferritin) if not performed in the past 90 days

D. PERFORM TARGETED EVALUATION

Evaluate if patient experiencing their typical VOE symptoms:

- ☐ If yes, confirm usual analgesic type and dose with patient

- ☐ If O_2 sat <95%, provide oxygen (not indicated if O_2 sat ≥95%)

If concerned for **PE**:

- ☐ Confirm adequate renal function
- ☐ Order CT angiogram

If concerned for **MI**:

- ☐ Order EKG
- ☐ Send troponin

If concerned for **acute chest syndrome**:

- ☐ Order chest radiograph
- ☐ Hematology consult, consider:
  - ☐ Adding hemoglobin fractionation/ electrophoresis to labs
  - ☐ Exchange transfusion

If concerned for **stroke** (e.g., patient reports headache):

- ☐ Implement SCD stroke protocol, if available
- ☐ Order brain imaging
- ☐ Hematology consult, consider:
  - ☐ Adding hemoglobin fractionation/ electrophoresis to labs
  - ☐ Exchange transfusion

If concerned for worsening **anemia**:

- ☐ Notify blood bank for phenotype-matched red cells

Add the following to labs:

- ☐ CBC with differential
- ☐ Type and screen
- ☐ Hemoglobin fractionation/electrophoresis
- ☐ Reticulocyte count
- ☐ LDH
- ☐ Total and direct bilirubin
- ☐ Iron studies (Fe, TIBC, Ferritin)

If concerned for **sequestration** or **acute cholecystitis** (e.g., patient reports abdominal pain):

- ☐ Order abdominal ultrasound

Add the following to labs:

- ☐ CBC with differential
- ☐ Type and screen
- ☐ ALT and AST
- ☐ Total and direct bilirubin

Assess **SIRS criteria**: T >38°C (100.4°F) or <36°C (96.8°F); HR >90; RR >20 or $PaCO_2$ <32mmHg; WBC >12,000/mm³, <4,000/mm³, or >10% bands

If ≥2 SIRS criteria present:

- ☐ Implement sepsis protocol
- ☐ Consider empiric treatment

E. INITIAL PAIN MANAGEMENT: Initiate analgesic therapy within 30 minutes of triage

- ☐ **If the patient has a documented individualized SCD pain plan, integrate here**
- ☐ If opioid is administered, initiate continuous O_2 sat monitoring

If the patient has an **opioid allergy**, provide alternative (assess renal/liver function as needed):

For children:

- ☐ PO acetaminophen 15mg/kg
- ☐ PO ibuprofen 10mg/kg
- ☐ If >2 years IV ketorolac 0.5mg/kg
- ☐ If <2 years IV ketorolac 0.25mg/kg

For adults:

- ☐ PO acetaminophen 975mg
- ☐ PO ibuprofen 600mg
- ☐ IV ketorolac 30mg

Alternative for adults:

- ☐ PO acetaminophen 650mg†
- ☐ IV ketamine 0.25mg/kg†
- ☐ IV ketorolac 15mg

If patient **has IV access** (e.g., peripheral or central line), administer IV opioid (1st dose):†

For patients who are **not** opioid naïve:

- ☐ Calculate and administer patient-specific opioid dose (IV route preferred)[ii]

For patients who **are** opioid naïve or with no available analgesic history, administer:

- ☐ Morphine 0.1mg/kg[iii]†
- ☐ Hydromorphone 0.02mg/kg[4]†
- ☐ Ketamine 0.25mg/kg[iv]†

Avoid meperidine†

If patient **does not have IV access**, administer opioid via other routes (1st dose):

For patients who are **not** opioid naïve:

- Calculate and administer patient-specific opioid dose (SQ if no IV access)[2]

For patients who **are** opioid naïve or with no available analgesic history, administer:

For children:

- PO hydromorphone 0.05mg/kg
- PO morphine 0.3mg/kg

Avoid IN fentanyl in patients under <7 years old or <10kg[5]

For adults:

- SQ morphine 0.1mg/kg
- SQ hydromorphone 0.02mg/kg
- PO morphine 30mg†
- PO hydromorphone 5mg
- IN fentanyl 2-3 doses 5 minutes apart (max single dose [100µg] may limit efficacy, especially >65kg)[v]

Consider adjunctive NSAIDs or acetaminophen (assess renal/liver function as needed):

For children:

- PO acetaminophen 15mg/kg
- PO ibuprofen 10mg/kg
- If >2 years IV ketorolac 0.5mg/kg
- If <2 years IV ketorolac 0.25mg/kg

For adults:

- PO acetaminophen 975mg
- IV ketorolac 15mg[vi]

Alternative for adults:

- PO ibuprofen 600mg†
- IV ketorolac 30mg†

If initial VAS ≥5:

Assess need for hydration (caution with CHF, pulmonary hypertension, or renal failure) and treat as needed

Assess itching:

- If itching in children: PO diphenhydramine 1mg/kg
- If itching in adults: PO diphenhydramine 25-50mg

Avoid IV diphenhydramine except in cases of anaphylaxis

Assess nausea:

- If nausea in children: IV ondansetron 4mg for >40kg, 0.1mg/kg for <40kg (PO if no IV)
- If nausea in adults: IV ondansetron 8mg (PO if no IV)

Note: IV magnesium and corticosteroids not indicated for SCD treatment†

F. FIRST PAIN REASSESSMENT: Within 30 minutes (60 minutes after triage)

Assess pain using VAS or verbal scale (1-10):

If VAS ≥5:

If no hypoxia or sedation:

- Repeat initial dose of IV opioid (2nd dose) if route is available (if route is not available, consider other routes)
- Escalate initial dose of IV opioid by 25%

If signs of excessive sedation:

- Decrease dose of IV opioid

If VAS ≤4 see **G [SECOND PAIN REASSESSMENT]**

G. SECOND PAIN REASSESSMENT: Within 30 minutes (90 minutes after triage)

- ☐ Assess vitals
- ☐ Perform follow-up lab tests† or review lab results and address abnormalities
- ☐ Re-evaluate for serious complications (see **D [TARGETED EVALUATION]**)

Assess pain using VAS or verbal scale (1-10):

If VAS ≥7:

If no hypoxia or sedation:

- ☐ Repeat 2nd dose IV opioid (3rd dose) if route is available (if route is not available, consider other routes)
- ☐ Escalate 2nd dose of IV opioid by 25%

If signs of excessive sedation:

- ☐ Decrease dose of IV opioid

Consider adjunctive NSAIDs or acetaminophen (assess renal/liver function as needed):

For children:

- ☐ PO acetaminophen 15mg/kg
- ☐ PO ibuprofen 10mg/kg
- ☐ If >2 years IV ketorolac 0.5mg/kg
- ☐ If <2 years IV ketorolac 0.25mg/kg

For adults:

- ☐ PO acetaminophen 975mg
- ☐ PO ibuprofen 600mg
- ☐ IV ketorolac 15mg[vi]

Alternative for adults:

- ☐ PO acetaminophen 650mg†
- ☐ IV ketorolac 30mg†

☐ Initiate PCA†
☐ Call for pain service consult
☐ Call for hematology/SCD expert consult
☐ Admit patient

If VAS 5-7:

If no hypoxia or sedation:

- ☐ Repeat 2nd dose IV opioid (3rd dose) if route is available (if route is not available, consider other routes)
- ☐ Escalate 2nd dose of IV opioid by 25%
- ☐ Consider switching opioid†

If signs of excessive sedation:

- ☐ Decrease dose of IV opioid

If VAS ≤4:

- ☐ Offer short-acting oral opioid

Assess if long-acting oral pain med prescribed as outpatient:

- ☐ If yes, restart long-acting oral pain med
- ☐ If no, call for pain service consult or SCD clinician team†

Ready for discharge (see **J [DISCHARGE]**):

- ☐ Call hematology/SCD expert about patient being readied for discharge†

H. THIRD PAIN REASSESSMENT: Within 30 minutes (120 minutes after triage)

- ☐ Assess vitals
- ☐ Review lab results and address abnormalities, including iron overload

Assess pain using VAS or verbal scale (1-10):

If VAS ≥5:

- ☐ Initiate PCA†
- ☐ Call for pain service consult
- ☐ Admit patient

If VAS ≤4 see **G [SECOND PAIN REASSESSMENT]**

I. PREVENTIVE CARE

Consider vaccinations:

- ☐ Consult CDC vaccination schedules[vii]
- ☐ If under age 5, twice-daily prophylactic penicillin†
- ☐ Pneumovax (wait ≥8 weeks since prior Prevnar)†

Inquire about access to behavioral health/psychiatric services:	Consult Case Management and social work:
☐ Order psychiatric referral†	☐ Support enrollment in appropriate services (e.g., disability)

J. DISCHARGE

- ☐ Confirm patient's pain is adequately controlled
- ☐ Schedule outpatient follow-up with PCP, hematology, or other SCD expert within 1 week

Determine the patient's current supply of pain medication:

If patient has adequate supply of pain medication, do not prescribe	If patient does not have adequate supply of pain medication:
	☐ Check prescription monitoring program (I-STOP)[viii] ☐ Prescribe 3-day supply of opioids† (consider 5 to 7-day supply) ☐ Prescribe adjunctive NSAIDs (consider renal function; should not be prescribed alone) ☐ Prescribe constipation prophylaxis

Provide and review SCD Pain Home Management discharge instructions and SCD education:

- ☐ Discuss signs of serious complications and instruct patient to return to ED if experienced (e.g., acute chest syndrome, stroke, sepsis, fever, etc.)
- ☐ Discuss addiction awareness
- ☐ Discuss overdose signs
- ☐ Prescribe Naloxone kits (for self and family members) if receiving ≥50 mg/day morphine equivalent dose
- ☐ Consider recommending that the patient discusses other disease-modifying treatments with hematologist:
  - ☐ Hydroxyurea
  - ☐ L-glutamine[ix]†
- ☐ Discuss setting up individualized treatment plan with SCD clinician

K. OTHER CONSIDERATIONS

Use non-pharmacologic approaches such as:

- ☐ Heat and distraction (e.g., music)
- ☐ Massage, yoga, TENS, virtual reality and guided audiovisual relaxation

†The majority of experts felt including this item would be likely to improve outcomes, but at least 2 experts disagreed.

ACRONYMS

ALT	Alanine aminotransferase
AST	Aspartate aminotransferase
CBC	Complete blood count
CDC	Centers for Disease Control and Prevention
CHF	Congestive heart failure
CT	Computerized tomography
ED	Emergency department
EKG	Electrocardiogram
ESI	Emergency severity index
HR	Heart rate
I-STOP	Internet system for tracking over-prescribing
IN	Intranasal
IV	Intravenous
LDH	Lactate dehydrogenase
MI	Myocardial infarction
NSAIDs	Nonsteroidal anti-inflammatory drugs
O_2 sat	Oxygen saturation
$PaCO_2$	Partial pressure of carbon dioxide
PCA	Patient-controlled analgesia
PCP	Primary care physician
PE	Pulmonary embolism
PO	Per os (oral)
RR	Respiratory rate
SCD	Sickle cell disease
SIRS	Systemic inflammatory response syndrome
SQ	Subcutaneous
T	Temperature
TIBC	Total iron-binding capacity
TENS	Transcutaneous electrical nerve stimulation
VAS	Visual analogue scale
VOE	Vaso-occlusive episode (sometimes referred to as vaso-occlusive crisis [VOC])
WBC	White blood cell count

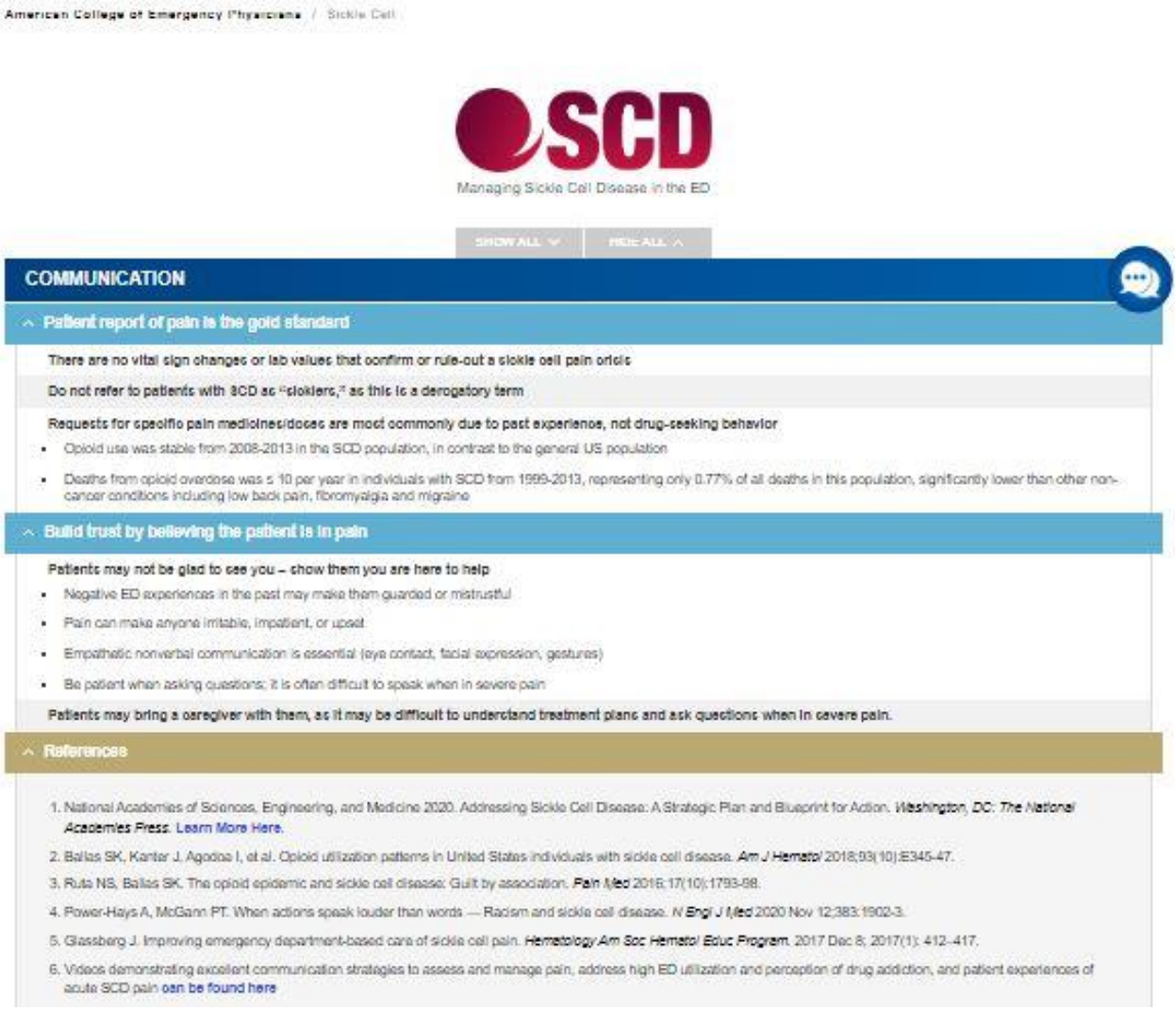
ADDITIONAL REFERENCES

[i] Brandow AM, DeBaun MR. Key Components of Pain Management for Children and Adults with Sickle Cell Disease. *Hematol Oncol Clin North Am*. 2018;32(3):535-550. doi:10.1016/j.hoc.2018.01.014
[ii] From Tanabe P, Silva S, Bosworth HB, et al. A randomized controlled trial comparing two vaso-occlusive episode (VOE) protocols in sickle cell disease (SCD). *Am J Hematol*. 2018;93(2):159-168. doi:10.1002/ajh.24948:

1. Review medical record for maximum home opioid dose and previous ED analgesic medication(s) and doses.
2. Calculate maximum home opioid dose by combining all long-acting and short-acting opioids taken within a 24-hour period.
    a. Suggested starting dose of IV opioid will be 20% of maximum home opioid dose, converted to IV morphine or IV hydromorphone.
    b. Suggested starting dose of oral opioid will be 20% of maximum home opioid dose, converted to any immediate release oral opioid that has worked for the patient in the past.
3. Compare the calculated dose to doses of opioids previously administered to the patient in the ED.
    a. If the ED doses and the calculated doses are within 10% of each other, keep calculated dose.
    b. If the discrepancy is greater than 10%, use the lower dose.

[iii] Tanabe et al. *Am J Hematol.* 2018.
[iv] Lubega FA, DeSilva MS, Munube D, et al. Low dose ketamine versus morphine for acute severe vaso occlusive pain in children: a randomized controlled trial. *Scand J Pain*. 2018;18:19-27.
[v] Kavanagh PL, Sprinz PG, Wolfgang TL, et al. Improving the Management of Vaso-Occlusive Episodes in the Pediatric Emergency Department. *PEDIATRICS*. 2015;136(4):e1016-e1025. doi:10.1542/peds.2014-3470
[vi] Beiter JL, Simon HK, Chambliss CR, et al. Intravenous ketorolac in the emergency department management of sickle cell pain and predictors of its effectiveness. *Arch Pediatr Adolesc Med*. 2001;155:496-500.
[vii] Available at https://www.cdc.gov/vaccines/schedules.
[viii] New York State Department of Health, Prescription Monitoring Program Registry (available at https://www.health.ny.gov/professionals/narcotic/prescription_monitoring/).
[ix] Niihara Y, Miller ST, Kanter J, et al. A Phase 3 Trial of l-Glutamine in Sickle Cell Disease. *N Engl J Med*. 2018;379:226-235.

American College of Emergency Physicians
Point-of-Care Tool: Sickle Cell Disease



American College of Emergency Physicians / Sickle Cell

SCD

Managing Sickle Cell Disease in the ED

SHOW ALL HIDE ALL

COMMUNICATION

Patient report of pain is the gold standard

There are no vital sign changes or lab values that confirm or rule-out a sickle cell pain crisis

Do not refer to patients with SCD as "sicklers," as this is a derogatory term

Requests for specific pain medicines/doses are most commonly due to past experience, not drug-seeking behavior

- Opioid use was stable from 2008-2013 in the SCD population, in contrast to the general US population
- Deaths from opioid overdose was ≤ 10 per year in individuals with SCD from 1999-2013, representing only 0.77% of all deaths in this population, significantly lower than other non-cancer conditions including low back pain, fibromyalgia and migraine

Build trust by believing the patient is in pain

Patients may not be glad to see you – show them you are here to help

- Negative ED experiences in the past may make them guarded or mistrustful
- Pain can make anyone irritable, impatient, or upset
- Empathetic nonverbal communication is essential (eye contact, facial expression, gestures)
- Be patient when asking questions; it is often difficult to speak when in severe pain

Patients may bring a caregiver with them, as it may be difficult to understand treatment plans and ask questions when in severe pain.

References

1. National Academies of Sciences, Engineering, and Medicine 2020. Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. *Washington, DC: The National Academies Press.* Learn More Here.
2. Ballas SK, Kanter J, Agodoa I, et al. Opioid utilization patterns in United States individuals with sickle cell disease. *Am J Hematol* 2018;93(10):E345-47.
3. Ruta NS, Ballas SK. The opioid epidemic and sickle cell disease: Guilt by association. *Pain Med* 2016;17(10):1793-98.
4. Power-Hays A, McGann PT. When actions speak louder than words — Racism and sickle cell disease. *N Engl J Med* 2020 Nov 12;383:1902-3.
5. Glassberg J. Improving emergency department-based care of sickle cell pain. *Hematology Am Soc Hematol Educ Program.* 2017 Dec 8; 2017(1): 412–417.
6. Videos demonstrating excellent communication strategies to assess and manage pain, address high ED utilization and perception of drug addiction, and patient experiences of acute SCD pain can be found here

TRIAGE

Sickle cell pain is usually severe and requires immediate treatment; evidence-based guidelines recommend administering pain medication within 60 minutes of arrival

Assign an emergent priority (e.g., ESI level 2) due to

- severe pain that cannot be managed in the waiting room
- high risk situation that needs rapid evaluation for other serious complications

Increase level of concern if pain is not similar to prior episodes or new symptoms are present

Patients with ≥ 3 admissions/year for sickle cell pain are at increased risk of death

Vital Signs/Neuro Status

Normal vital signs do not rule out pain crisis

Patient report of pain is gold standard

Hypoxia should be judged against patient's baseline

Fever ≥ 100.4 degrees (≥ 38 degrees C) should prompt work-up for underlying cause, as sickle cell disease decreases splenic function

- Children and adolescents with fever ≥ 101.5 degrees F (≥ 38.5 degrees C) should receive broad spectrum antibiotics ≤ 1 hour of arrival

Hypotension and tachycardia should raise concern for sepsis, dehydration and significant anemia due to aplastic anemia or splenic sequestration

Tachypnea should raise concern for acute chest syndrome, occult hypoxemia and pulmonary embolus

Altered mental status, severe or atypical headache, focal neurologic findings (such as vision changes), or new seizure should raise concern for stroke in both children and adults

Past Medical History

Ask about history of severe SCD complications (such as acute chest syndrome, pain crises, stroke), recent admissions and recent blood transfusions

Patients are born with SCD; onset and severity of symptoms vary

Initiation of Care Should Not Be Delayed Due to Space Constraints

Consider alternative spaces (such as asthma bay) if no treatment spaces available

Consider administering pain medications in triage (e.g., subcutaneous opioids for adults and intranasal fentanyl for children – see Treatment)

Utilize physician in waiting room, split flow model, etc. to expedite care when available

References

1. U.S. Department of Health and Human Services. National Heart Lung, and Blood Institute. Evidence-Based Management of Sickle Cell Disease, Expert Panel Report, 2014 - View PDF.
2. National Academies of Sciences, Engineering, and Medicine 2020. Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. Washington, DC: The National Academies Press. Learn more here.
3. Elmariah H, Garrett ME, de Castro LM, et al. Factors associated with survival in a contemporary adult sickle cell cohort. *Am J Hematol* 2014;89(5):530-35.
4. Videos demonstrating excellent communication strategies to assess and manage pain, address high ED utilization and perception of drug addiction, and patient experiences of acute SCD pain can be found here.

HISTORY

History – Part 1

From the patient

- Where is pain most intense? Where does it hurt the most?
- Is there anything unusual or concerning about the pain you are experiencing?
  - Does this feel like your typical pain crisis?
  - If the pain is different, how is it different – in location? Sensation? Intensity?
- Have you had a fever in the last 24 hours?

From patient, caregiver, or chart (patient may be in too much pain to give complete history)

- When did the pain start?
- What medicines did you try at home? What dose did you take, and when? Did they help?
- Do you have a pain plan in your chart (with preferred pain medicines and doses)? If not:
  - What pain medicine(s) work best for your pain in the ED? Do you know what dose works for you?
  - Are there any pain medicines that do not work, or you have had a bad experience with?
- What other things will make you more comfortable?
  - Heat packs
  - Blankets
  - Distraction – such as talking on the phone or watching TV

TIP: Share the initial plan and rationale with the patient and caregiver (if present)

History – Part 2 (after Evaluation and initial orders placed)

From chart, patient or caregiver (patients may be in too much pain to give complete history)

- Do you have pain most days? What level of pain is typical for you?
- What do you take every day for pain?
- If patient is followed at your institution:
  - I have your list of medicines here (review list); are there others that you take?
  - I see that you have had the following problems in the past (review list); are there others that I missed?
  - I found/did not find (list) surgeries in your chart (review list), are there others that I missed?
- If patient is not followed at your institution:
  - Do you have a primary hospital, clinic or doctor who treats you for sickle cell disease? Do you have other doctors that you see, such as primary care?
  - Do you take medicines every day, such as hydroxyurea? Are there other medicines you take only when needed (such as for pain)?
  - When was your last blood transfusion? Do you get regularly scheduled blood transfusions (~once/month)?
  - Do you have any SCD-related health issues?
    - History of acute chest syndrome, stroke, sepsis, kidney problems, blood clots?
    - Have you had surgery, such as removal of gall bladder or spleen?
  - What is your hemoglobin or hematocrit when you are well?

TIP: Regularly update the plan and rationale with the patient and caregiver (if present)

References

1. U.S. Department of Health and Human Services. National Heart Lung, and Blood Institute. Evidence-Based Management of Sickle Cell Disease, Expert Panel Report, 2014. View PDF here.
2. National Academies of Sciences, Engineering, and Medicine 2020. Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. Washington, DC: The National Academies Press. Learn more here.
3. Videos demonstrating excellent communication strategies to assess and manage pain, address high ED utilization and perception of drug addiction, and patient experiences of acute SCD pain can be found here.

EVALUATION

Vital Signs

Normal vital signs do not rule out pain crisis or correlate with severity of pain

Hypoxia should be judged against patient baseline; raises concern for acute chest, pneumonia, and pulmonary embolism

Tachypnea should raise concern for acute chest syndrome and pulmonary embolism

Fever should prompt rapid work-up for underlying cause since patients are functionally asplenic; also consider delayed hemolytic transfusion reaction if transfused in previous 4 weeks

Hypotension and tachycardia raise concern for sepsis, dehydration and significant anemia due to aplastic crisis or splenic sequestration

Physical Exam/Differential Diagnosis

NOTE: Severe SCD complications often develop during a pain crisis or mimic one. Contact hematology EMERGENTLY if you suspect one, such as acute stroke or acute chest syndrome with significant respiratory distress.

Location of pain

- Localized bony pain that is different from previous pain crises
  - Osteomyelitis/septic arthritis
  - Priapism (sometimes report thigh pain)
  - Avascular necrosis
- Calf pain could be DVT
- Diffuse pain could be systemic infection: Check vital signs, look for rash or petechiae on skin (including palms/soles) and gums

Altered mental status, severe or atypical headache, focal neurologic findings (such as vision changes), or new seizure: Consider stroke in children and adults

Fatigue/jaundice/pallor: Consider aplastic anemia, sepsis or hemolysis due to delayed hemolytic transfusion reaction

Chest pain: Consider acute chest syndrome (leading cause of death in SCD)

Respiratory distress: Consider acute chest syndrome, pulmonary embolism and sepsis

Abdominal pain or distension: Assess for splenomegaly or hepatomegaly (splenic or hepatic sequestration), RUQ pain (acute cholecystitis), and consider priapism if low abdominal pain

Assess fluid status for dehydration

Laboratory Workup

Use order set for SCD if available

Get CBC with differential and reticulocyte count; consider comprehensive metabolic panel in adults if clinically indicated.

- Hemoglobin and retic count should be interpreted relative to baseline values
  - Anemia + low retic = concern for aplastic anemia
  - Anemia + low platelet count + normal retic = concern for splenic sequestration
- Anemia can progress extremely quickly due to low baseline hemoglobin and short half-life of RBCs due to clearance of abnormal cells.

Obtain blood culture in all children with fever, and adults with central venous access or history of sepsis or splenectomy

Imaging

CXR (2 view if able) for fever, chest pain, or respiratory symptoms/findings to assess for acute chest syndrome

Other Considerations

Patients should not be penalized for using coping mechanisms to manage pain, including naps, distraction techniques, discussions with family, or meditation

Patients with frequent visits to the ED are at increased risk of serious complications

- Patients with ≥ 3 admissions/year for sickle cell pain are at increased risk of death
- Some may also have significant mental health and/or psychosocial complications which can exacerbate their disease course. This should not be used to dismiss the severity of their pain. Consult social work or case management if available.

References

1. U.S. Department of Health and Human Services. National Heart Lung, and Blood Institute. Evidence-Based Management of Sickle Cell Disease, Expert Panel Report, 2014. View PDF here.
2. National Academies of Sciences, Engineering, and Medicine 2020. Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action. Washington, DC: *The National Academies Press*. Learn more here.
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4. Elmariah H, Garrett ME, de Castro LM, et al. Factors associated with survival in a contemporary adult sickle cell cohort. *Am J Hematol* 2014;89(5):530-35.

TREATMENT

Treat Immediately

Sickle cell pain is usually severe and requires immediate treatment

Do not wait for lab results before starting pain medications; there are no lab values that confirm or rule-out a sickle cell pain crisis

Requests for pain medicines/doses are most commonly due to past experience, not drug-seeking behavior

- Opioid use was stable from 2008-2013 among individuals with SCD, in contrast to the general US population
- Deaths from opioids was ≤ 10 per year in individuals with SCD from 1999-2013 (representing only 0.77% of deaths in this population), significantly lower than other non-cancer conditions including low back pain, fibromyalgia and migraine

Evaluation and treatment should begin in alternative spaces if no treatment rooms available

Consider administering pain medicines while in waiting room/triage

Treatment Approach

Use a standardized pain plan or protocol to treat acute SCD pain

Check electronic health record for an existing patient-specific pain plan (individualized medications and doses); if none found, elicit patient preference based on previous experiences.

Treating acute moderate to severe pain

- **1st Dose within 60 min of ED arrival**
  - Consider intranasal fentanyl as 1st dose in children
  - Consider subcutaneous route for adults with difficult IV access
- **Reassess & re-dose every 30 min**
  - until pain improves
- **Reassess 15-30 min after 1st dose**
  - Can increase dose by 25% once if no change

Routes of medication administration

- Use ultrasound-guided IV placement if available; gaining IV access is often challenging
- Subcutaneous route can also be used to provide rapid analgesia
- Avoid IM doses of opioids unless other routes attempted and failed

Sleep following opioids does not usually indicate pain control

- Wake the patient, assess pain level and re-dose based on patient report
- Drowsiness is a side effect of opioids, compounded by sleeplessness due to severe pain

Suggested Parenteral Opioid and NSAID Doses for Children (< 12 years or < 60 kg):

Pain Medication	Dose	Max Single Dose	Frequency
Intranasal fentanyl*	2 mcg/kg	100 mcg or 1 ml per nare	May repeat x1 after 10 minutes
Morphine IV**	0.1 mg/kg	10 mg	Repeat every 15-30 minutes until pain controlled
Hydromorphone IV	0.015 mg/kg	1.2 mg	Repeat every 15-30 minutes until pain controlled
Ketorolac IV**	0.5 mg/kg	15-30mg	Once

*Recommended as 1st pain medication in children
**Use with caution in patients with mild renal dysfunction

Suggested Parenteral Opioid and NSAID Doses for Adults and Adolescents (≥ 12 years or ≥ 60 kg)

Pain Medication	Dose	Max Single Dose	Frequency
Morphine IV*	0.1 mg/kg	10 mg	Repeat every 15-30 minutes until pain controlled
Hydromorphone IV	0.015 mg/kg	4 mg	Repeat every 15-30 minutes until pain controlled
Ketorolac IV*	0.5 mg/kg	15 mg	Once

*Use with caution in patients with mild renal dysfunction

TREATMENT

Treat Immediately

Treatment Approach

Adjuncts for Pain

NSAIDs: Can be given in addition to opioids to manage acute pain and treat the inflammation associated with a sickle cell crisis

- Exclusions include patients with significant renal dysfunction (up to 30% of adults with SCD) and full-dose anticoagulation; use caution in patients with peptic ulcer disease

IV fluids: No evidence that IV fluids are beneficial in euvolemic patients; may cause fluid overload and acute chest syndrome in certain patients

- Consider IVF infusion rather than bolus and administer less than maintenance fluids if able to drink (e.g., 75% maintenance)

Non-pharmacologic interventions: Heat packs (no ice), blankets, distraction (using phone, watching TV)

Manage Opioid Side Effects

Pruritus:

- Oral diphenhydramine 25 mg recommended. Avoid IV diphenhydramine push because it worsens sedation.
- Low dose naloxone infusion (1 mcg/kg/hr) does not reverse effects of opioids, can be titrated up 0.5 mcg/kg/hr every 3-4 hours, maximum 4-5 mcg/kg/hr
- IV diphenhydramine 25 mg if alternatives listed above do not control pruritus or are unavailable

Nausea/vomiting: Ondansetron is commonly used; use caution with prochlorperazine and metoclopramide due to sedating effects

Respiratory depression: Naloxone 0.4 mg/dose IV to alleviate sedation (not reverse analgesia)

Patient-Controlled Analgesia (PCA)

Consider for patients being admitted to prevent lapses in dosing

If EHR order set, pharmacy/hospital protocols, and/or SCD patient plans are available, use demand and basal dosing regimens; otherwise use demand dosing alone

Suggested Doses for Children (< 12 years or < 50 kg)

Medication	Demand Dose	1 Hour Limit
Morphine	0.015 - 0.02 mg/kg	0.12 mg/kg
Hydromorphone	0.003 mg/kg	0.03 mg/kg

Notes: Lockout period 8-10 minutes. Titrate as needed.

Suggested Doses for Adults and Adolescents (≥ 12 years or ≥ 50 kg)

Medication	Demand Dose	1 Hour Limit
Morphine	2 mg (0.02-0.03 mg/kg)	10 mg
Hydromorphone	0.4-1 mg (0.01-0.03 mg/kg)	4 mg

Notes: Lockout period 10 minutes. Titrate as needed. Order oral long-acting opioid if prescribed

Treatments to Use with Caution or Avoid

Steroids: Can trigger severe pain crises, but short courses can safely treat select conditions (e.g., asthma) when done in coordination with hematology/SCD provider

Ketamine: Guidelines found insufficient evidence for use outside of the inpatient setting

Blood transfusions: Do not give transfusions to manage sickle cell pain crisis unless directed by the patient's hematologist or SCD provider

Oxygen: Do not use oxygen unless patient is hypoxic compared to their usual oxygen level

Special Populations

Pregnant patients with pain crisis require treatment with opioids; the benefits of treatment outweigh the harms to fetus. Offer acetaminophen as an option or adjunct for pain

For patients on methadone, long-acting opioids, or lidocaine or fentanyl patch: Continue home dose

For patients on methadone or buprenorphine: Look for individualized pain plan; if none found, consult hematologist/SCD provider for guidance on pain medication dosing

TREATMENT

- Treat Immediately
- Treatment Approach
- Adjuncts for Pain
- Manage Opioid Side Effects
- Patient-Controlled Analgesia (PCA)
- Treatments to Use with Caution or Avoid
- Special Populations
- References

1. Brandow AM, Carroll CP, Creary S, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. *Blood Advances*. 2020;4(12):2656-2701.
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5. He F, Jiang Y, Li L. The effect of naloxone treatment on opioid-induced side effects: A meta-analysis of randomized and controlled trials. *Medicine* 2016;95(37):e4729. doi: 10.1097/MD.0000000000004729.
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DISPOSITION

Admit/Observe

Consider admission if pain is not well-controlled; patients usually know if their pain has stabilized at a manageable level

Other complications found (e.g., infiltrate on CXR; suspect serious or systemic infection)

Consider admission for young (< 1 year) patients, or older patients with fever if they cannot seek care quickly or follow-up with SCD provider the next day

Consider alternative treatment settings for improving but not relieved pain, such as observation units, to allow for a longer duration to treat pain and spare a hospital admission

Discharge

If patient believes they can manage pain at home

Recommend close follow-up with hematology/SCD provider. Refer to a specialist if not already established

- Recognize that there is not always ready access to a hematologist. Efforts should be made to establish a referral pathway to a local physician to manage these patients

Ensure adequate access to medications for management of pain as an outpatient

- o Opioids:
  - Short-term (e.g., 3 days) opioid prescription as bridge to next outpatient visit
  - If concern for opioid misuse/overdose – contact outpatient provider to make plan
- Non-opioids including acetaminophen, topical or oral NSAID (if not contraindicated): Continue for short-course (5-7 days total)
- Non-pharmacologic means of pain control: Continue heat, hydration, distraction; avoid using ice
- Bowel regimen to avoid opioid-induced constipation

Give clear return precautions, such as fever, difficulty breathing, chest pain, changes in mental status, uncontrolled pain

Some patients with frequent visits have unmet social or behavioral health needs including lack of insurance or transportation, unable to pay for prescriptions, unstable housing, etc. Make referrals to social work or case management as appropriate. Learn more here.

References

1. Brandow AM, Carroll CP, Creary S, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. *Blood Advances*. 2020;4(12):2656-2701.
2. U.S. Department of Health and Human Services. National Heart Lung, and Blood Institute. Evidence-Based Management of Sickle Cell Disease, Expert Panel Report, 2014. View PDF here.
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