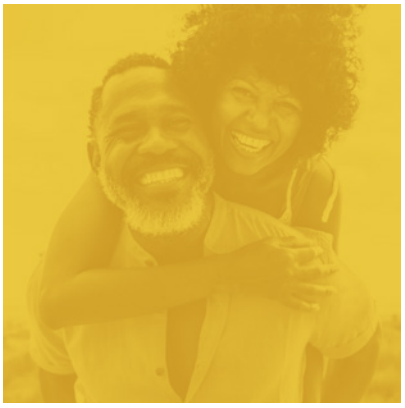
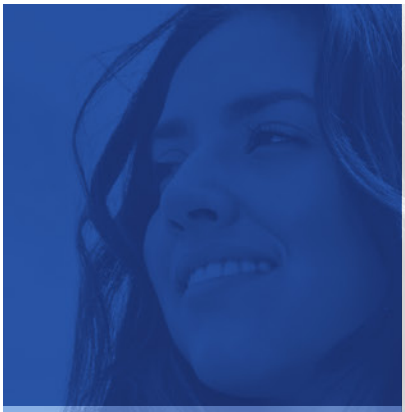




CMS Sickle Cell Disease Provider Toolkit

RESOURCES FOR HEALTH CARE PROFESSIONALS TO SUPPORT INDIVIDUALS WITH SICKLE CELL DISEASE

Contents

CMS Sickle Cell Disease (SCD) Health Care Provider Toolkit: Resources for Health Care Professionals to Support Individuals with SCD

Background

Building on the CMS SCD Action Plan launched in September 2023, this toolkit is intended to assist health care providers and the care team supporting individuals with SCD in primary care and other settings. Members of the care team can use this toolkit to understand the basics of SCD, including information on screening, diagnosis, and treatment, as well as how to identify resources to support individuals with SCD. The toolkit also provides information on how CMS program coverage can assist people with SCD and educational materials for individuals with SCD and other community partners who serve them.

1. Screening and Diagnosis

Sickle cell disease (SCD) is an autosomal recessive inherited disease is considered "rare", but nonetheless affects an estimated 100,000 Americans (the majority being of African descent) and 1 in 365 African American births.[1] The disease is caused by a point mutation in the beta-globin gene, which leads to polymerization of hemoglobin molecules which can create the characteristic sickle cell shape.[2]

SCD is a recessive genetic condition that resu[3]lts when a person inherits two copies (homozygote) of the beta-globin gene containing a point mutation that changes one of the amino acids in the protein. The resulting hemoglobin with this variant is called hemoglobin S. A person who inherits only one copy (heterozygote) of this variant and one unmutated copy is said to have sickle cell trait. While these individuals can pass this variant on to their offspring, sickle cell trait causes few to no medical issues in most carriers. The inheritance of two copies leads to a hemoglobin that forms strands in the absence of oxygen, ultimately leading to the adverse consequences seen in SCD.[3]

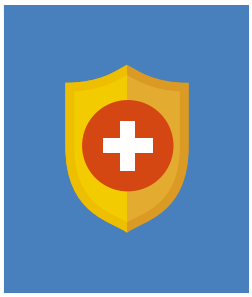
Health care providers may use prenatal screening, blood testing, or genetic testing to diagnose SCD. Prenatal screening can diagnose SCD as early as 8 to 10 weeks into a pregnancy. This screening uses either a sample of amniotic fluid or a sample of tissue from the placenta to test for the presence of the sickle cell hemoglobin gene. These tests cannot predict the severity of the disease.[4]

Individuals may be diagnosed with SCD through blood tests, most commonly performed during newborn screening. Newborn screening for SCD occurs in every state and is performed within the first 24 to 48 hours after birth as part of a larger panel.[5] The screening involves a simple heel prick and the collection of a few drops of blood which are tested for a variety of congenital disorders.[6] This blood test can also determine whether the newborn carries the sickle cell trait.

Genetic testing can aid in determining what type of SCD an individual has and can help confirm a diagnosis if the results from blood tests are unclear. Genetic counseling is also an essential part in minimizing the burden of SCD and ensuring that information is clearly communicated.[7,8]

For more information on screening and diagnosis for SCD, see:

- Sickle Cell Disease Diagnosis (NIH)
- Hemoglobinopathies: Current Practices for Screening, Confirmation and Follow-up (CDC)
- Newborn Screening for Providers (HRSA)



2. Treatment

Medical Treatments & Medications

Hydroxyurea was the first medication approved by the U.S. Food and Drug Administration (FDA) to treat sickle cell and is still used as first-line treatment.[9] The oral medication has been shown to reduce sickling and reduce or prevent several complications of SCD. If hydroxyurea does not control symptoms enough, the newer FDA-approved drugs may be added on top of hydroxyurea treatment for combination therapy.[10]

Other common treatments for SCD often aim to prevent or minimize complications associated with SCD. These include hydroxyurea to reduce or prevent complications of SCD, and penicillin to reduce risk of infection in children, crizanlizumab-tmca or L-glutamine glutamine to reduce pain crises. To manage acute pain that arises from vaso-occlusive episodes, health care providers may recommend over-the-counter pain medication, non-opioid or opioid pain medication, and/or adjuvant therapies (e.g., breathing exercises), depending on severity. To manage chronic pain that occurs in most people with SCD, an integrated care approach of medication and non-pharmacologic treatments may be recommended.

Healthcare providers may recommend a blood transfusion to treat and prevent certain sickle cell disease complications, including for children who have abnormal transcranial Doppler ultrasound results because transfusions can lower the chance of having a first stroke. Transfusions may also help lower the chances of secondary stroke in adults who have had an acute stroke.[11]

For additional information on clinical guidelines, best practices, and treatment for SCD, see:

- Sickle Cell Disease Clinical Guidelines (CDC)
  This resource provides links to clinical guidelines developed by the National Heart, Lung, and Blood Institute as well as additional CDC resources.
- Sickle Cell Disease Treatment (NIH)
- Hydroxyurea for the Treatment of Sickle Cell Disease (AHRQ)
- Hydroxyurea Use for Sickle Cell Disease Fact Sheet (NIH)
- Gene Therapy Educational Materials for the Sickle Cell Disease Community (NIH)

Transformative Sickle Cell Treatments: Blood and Bone Marrow Transplant & Gene Therapies

Until recently, a bone marrow transplant was the only cure for sickle cell disease.[12] However, in December 2023, the U.S. Food and Drug Administration (FDA) approved two new genetic therapies to treat the disease.[13] Even so, a bone marrow transplant is still a curative option, but it is not for everyone. To be successful, transplants require a genetically well-matched donor, usually a family member. Most transplants are performed in children and young adults who have had complications, such as strokes, acute chest syndrome, and recurring pain crises. Blood and bone marrow transplants are riskier in adults.[11]

Cell-based gene therapies are a growing class of transformative, typically one-time medicines designed to treat, or even potentially cure, previously intractable diseases. CMS and other federal agencies are committed to advancing policy solutions, including the CMS Innovation Center's Cell and Gene Therapy (CGT) Access Model, to support increased access to these new high-cost treatments. See Section 4 of this toolkit for more information on the CGT Access Model.

For more information on treatments and clinical trials, including those testing gene therapies for SCD, see:

- Cure Sickle Cell Initiative (NIH)
- Treatments for Blood Disorders (NIH)

3. Comprehensive Care for People with SCD

Health care professionals and community health workers (CHWs) play a vital role in identifying and providing education about the early symptoms of SCD, helping individuals manage health complications associated with SCD, and determining when urgent or emergency medical care is necessary for life threatening health concerns or severe acute pain episodes. This enables them to respond in a timely fashion, administer appropriate care, and provide education to patients with SCD and their caregivers and families. The Health Resources and Services Administration (HRSA) funds the Sickle Cell Disease Treatment Demonstration Program that works to improve

health care and outcomes for individuals with SCD. This program funds five comprehensive sickle cell disease centers that provide care to people living with SCD in their geographic region. They provide direct clinical care and help other clinics, through training, to deliver evidence-based care. The program is designed so that experts in centrally based hospitals, clinics, or university health centers (the "hub") provide help to the communities where people live and use services ("spokes").

Culturally Tailored Whole Person Care

Whole person care is a holistic approach in health care that looks at the whole person and considers all the factors that can influence health or disease.[14] Given the complexity and impact of SCD on patient quality of life and outcomes, health care providers should be mindful of a person's physical, mental, and emotional well-being when treating a patient with SCD.[15]

More specifically, whole person care is important to provide to people with SCD because it:

- Recognizes that managing SCD involves more than treating physical symptoms
- Considers the physical, emotional, social, spiritual and psychological needs of individuals with SCD
- Provides comprehensive and personalized support beyond treating symptoms or the disease
- Improves treatment adherence, reduces complications, and enhances health outcomes
- Helps reduce disparities in health care access and outcomes for communities impacted by SCD
- Prioritizes individual needs and goals, leading to better quality of life
- Addresses the pain experience in individuals with SCD requiring a multidisciplinary and multi-modal treatment approach[16]

SCD disproportionately impacts or African American and Hispanic or Latino individuals. Societal factors such as stigma associated with the disease and discrimination contribute to the burden of living with SCD.[17]People with SCD experience worse access to care and health outcomes compared to those with other diseases, exacerbating underlying inequities faced by these populations. Persistent and systemic inequities in our health care system have created barriers for these patients in receiving help.

The National Culturally and Linguistically Appropriate Services (CLAS) Standards are a set of recommended standards which aim to advance health equity, improve the quality of health care, and eliminate health disparities by providing a framework for health care providers and health care organizations to deliver services that are culturally and linguistically appropriate, respectful, and responsive to patient's cultural health beliefs, preferences, and communication needs.

Resources:

- Sickle Cell Disease: Tips for Healthy Living Fact Sheet
- Caring for Young Children with Sickle Cell Disease
- CHWs as Support for Sickle Cell Care
- A Practical Guide to Implementing the National CLAS Standards: For Racial, Ethnic and Linguistic Minorities, People with Disabilities and Sexual and Gender Minorities

Disparities in Access to Care for People with SCD in the U.S.

Wait Times: Patients with SCD experienced 25% longer wait times in the emergency department compared to other patients.[18]

Lack of Physicians: There is a shortage of hematologists trained to treat SCD. This shortage leads to an over-reliance on emergency care, especially for adults with SCD.[19] This is especially an issue for individuals with SCD in rural areas, where access to care can be especially limited.

Health Coverage: Many individuals with SCD experience barriers in accessing care, which results in delays in treatment.[20]

Barriers to Receiving Pain Treatment: Though guidelines recommend that patients receive their first dose of pain medication within 60 minutes of arriving to the emergency room for an acute pain episode, 50% of SCD patients reported waiting 2 hours or longer.[21]

Lack of Knowledge Among Health Care Providers: Individuals with SCD report their providers have a lack of knowledge, training, and understanding of SCD, including health care providers who prescribe pain medications only without understanding the full SCD pathology or SCD-specific treatments.[22] In addition, health professionals often view patients with SCD as “drug seeking”, which exacerbates stigma around the disease.[23]

Disparities in Health Outcomes in the U.S.

Life Expectancy: The lifespans of individuals with SCD are 20+ years shorter compared to the average life expectancy in the U.S. The median life expectancy for publicly insured individuals with SCD is an average of 52.6 years,[24] contrasting with average life expectancy in the U.S. of 77.5 years.[25]

However, in the past several years, new treatments and management strategies have increased life expectancy for people living with SCD, and an increasing number are living long enough to become age-eligible for Medicare.

Readmission Rates: Patients with SCD have the highest 30-day hospital readmission rates compared to other health conditions.[26]

Rate of Stroke: The risk of stroke for children with SCD is over 100x that of children without SCD.[27] In addition, the rate of stroke in adults (aged 35-64) with SCD is 3x higher than rates among African Americans of similar age without SCD.[26]

SCD Health Care Transition from Pediatric to Adult Care

SCD health care transition (HCT) is a planned process in which pediatric patients with SCD mature and shift from receiving pediatric care to adult care, with or without transferring to a new clinician. HCTs occur in three phases: transition preparation, transfer, and adult care integration.[28]

Why are health care transitions important for pediatric patients with SCD?

HCTs aim is to establish an organized approach in pediatric and adult care practices to promote transition readiness, care transfer, and integration into adult-centered care and to facilitate positive disease management as young adults learn to take care of their own health and wellbeing and mature into independent adults.[28]

Patients with SCD who experience a delay of over six months in transitioning from pediatric to adult care are twice as likely to be hospitalized compared to those who transition in less than two months.[29] In addition, pediatric individuals with SCD who do not successfully transition may experience worsening disease severity, and greater risk for early death.[30]

Transitions can be especially challenging for young people as they often co-occur with other major life changes like starting a career, beginning college, or moving to a new location and/or living independently.[31] In addition, for children in Medicaid/CHIP, program eligibility and benefits may change as children age, affecting access to services and supports. Therefore, a comprehensive and coordinated approach is needed to ensure a seamless transfer of medical records, treatment plans, and specialized care protocols among health care providers.

Pain Management

The pain trajectory in SCD is unique and complicated. There is enormous heterogeneity in SCD-related pain, as well as within-individual variability in pain phenotypes. People with SCD often describe pain as deep, stabbing, electrical, throbbing, beating, spreading, or feeling like broken glass in the veins. They may experience pain anywhere in their body, and they may endure acute pain, chronic pain, or both. Acute pain episodes (also known as vaso-occlusive episodes) occur when sickled cells obstruct blood flow as they travel through small blood vessels, which leads to intense pain. Forty-eight percent of individuals with SCD reported four or more acute pain episodes in the past 12 months.[32]

On average, individuals with SCD visit the hospital more than once a year and visit the emergency department two to three times a year. Acute pain episodes are the main driver of hospital stays and emergency department visits among those with SCD.[33] Due to racial bias and lack of training surrounding SCD, people with SCD commonly report that health professionals view them as drug seeking, which exacerbates stigma around the disease.[23]

Pain management for individuals with SCD is complex and presents unique challenges that must be considered. The goal is to reduce the burden of the presenting pain type (e.g., acute pain episode vs. chronic pain) on the person and improve quality of life. Healthcare providers may recommend treatments such as L-glutamine, crizanlizumab-tmca, and hydroxyurea. Common treatments for acute pain episodes include nonsteroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen, in mild cases, and opioids such as oxycodone, hydrocodone, and morphine.[11] While opioid therapy remains vital for the management of acute pain episodes, nonopioid pharmacologic and nonpharmacologic (complementary) approaches for chronic pain management have been deemed feasible and of interest to individuals with SCD. Chronic pain management is based on the biopsychosocial nature of pain. Care providers may recommend medication and complementary, nonpharmacologic approaches, such as mind-body interventions (e.g., acupuncture, yoga, meditation), cognitive-behavioral therapy, and dietary or nutritional modifications (e.g., special diets).[34] Often, an integrative approach is recommended, which combines complementary approaches with conventional medical interventions such as pharmacologic, surgery, or device-based treatments.

For more information on pain management, including resources for SCD pain management and resources for general chronic and acute pain management, see:

- Steps to Better Health Toolkit: Managing Chronic Pain (CDC)
- Steps to Better Health Toolkit: Managing Acute Pain (CDC)
- Managing Pain with Sickle Cell Disease Fact Sheet (NIH)
- CDC Clinical Practice Guideline for Prescribing Opioids for Pain (CDC)
  This guideline includes information about the co-prescription of opioids and naloxone. However, it excludes pain management related to SCD and recommends disease-specific guidelines for managing pain with SCD.

Telehealth

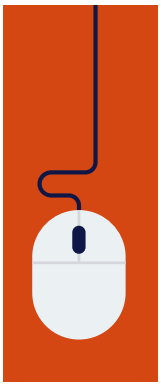
Telehealth, sometimes referred to as telemedicine, is the use of electronic information and telecommunications technologies to extend care when the provider and the patient aren't in the same place at the same time. Technologies for telehealth include videoconferencing, store-and-forward imaging, streaming media, and terrestrial and wireless communications.[35]

Telehealth is a promising practice for providing care to people with SCD, especially in rural regions and geographic locations where there may be a limited number of health care providers. During the COVID-19 pandemic, patients with SCD expressed positive experiences with telehealth and found it to be an effective way to receive treatment and reduce numerous obstacles to accessing care. They also reported interest in continuing telemedicine care after the COVID-19 pandemic.[36] In addition to improved access to care, other advantages of telehealth in rural communities include:[37]

- Reduced expenses for patients such as travel costs and time
- Decreased operational costs for health care providers like staffing
- Less utilization of onsite health care resources
- Stronger retention and recruitment of providers
- Education and training opportunities for patients and health care providers

For more information about telehealth, see the following CMS resources:

- Coverage to Care (C2C) Telehealth for Providers: What You Need to Know
  This resource includes information about telehealth, steps for using telehealth, considerations with various populations, and billing for telehealth.
- Medicare Telehealth resources
- Medicaid and CHIP Telehealth Toolkit



Supporting Social Determinants of Health for People with SCD

Social determinants of health (SDOH) are the conditions in the environment in which people are born, live, work, learn, play, worship, and age that impact a person's health, well-being, and overall quality of life. SDOH can include transportation, neighborhoods and safe housing, education and job opportunities, income, discrimination and violence, polluted air and water, and access to healthy foods and physical activity, and literacy skills.[38]

SDOH impact the treatment, management, and health outcomes associated with SCD, and overall quality of life for individuals with SCD. By understanding the broader context in which patients with SCD and their families live and the challenges they face, health care organizations and members of the care team can strive to reduce health disparities by addressing underlying social factors that impact health, such as transportation, nutrition, and housing. Health care organizations can help identify people's SDOH and health related social needs (HRSN) and

their risk for developing social needs through screening tools. HRSN are an individual's unmet adverse social conditions that contribute to poor health outcomes. Please see Section 4 below for more information about payment for services that may address SDOH.

For more information about social needs screenings, see:

- Tools To Help Health Care Organizations Address SDOH (AHRQ)
- Accountable Health Communities Health-Related Social Needs Screening Tool (CMS)
- Health Related Social Needs (Medicaid)
- Social Determinants of Health Information Exchange Toolkit (ONC)

4. CMS Programs and Resources that Support Comprehensive SCD Care

Across Medicaid, the Children's Health Insurance Program (CHIP), Medicare, and the Health Insurance Marketplace, there are varying mechanisms for payment for key treatments and supports for individuals with SCD.

Payment and Coverage for Primary Care

See this resource for primary care coverage under Medicare:

- Medicare Preventive Services (Medicare Learning Network)

Payment and Coverage for Medical Treatments

Medicaid and CHIP

Federal law outlines mandatory Medicaid and CHIP benefits, which states are required to cover, and optional benefits that states may cover if they choose. Mandatory Medicaid benefits for many beneficiaries include:

- Inpatient and outpatient hospital services;
- Laboratory and X-ray services; physician services;
- Early and periodic screening, diagnostic, and treatment services (EPSDT) for eligible beneficiaries under age 21, including well-baby and well-child visits;
- Medication Assisted Treatment (MAT) for opioid use disorder family planning services; and
- Access to transportation to medical care.

Some examples of optional benefits include:

- Diagnostic and screening services for adults;
- Preventive services;
- Rehabilitative services occupational and physical therapy; and
- Clinic services.

In separate CHIPs, all states must provide well-baby and well-child care, dental coverage, behavioral health care, and age-appropriate ACIP-approved vaccines. Under Medicaid, if a state determines, on the basis of medical advice, that needed medical services (e.g., gene therapy providers) are more readily available in another state, then the state must pay for services from the out-of-state provider to the same extent it would pay for services furnished in-state.[39]

States may also cover an optional benefit for certain SCD treatment and services.[40, 41] Outpatient prescription drug coverage generally is an optional benefit that all state Medicaid agencies currently provide.[42] State Medicaid agencies that provide this optional outpatient prescription drug coverage are required to cover all covered outpatient drugs offered by any manufacturer that agrees to provide rebates. Drugs that are administered in an inpatient hospital setting are considered covered outpatient drugs if they are directly reimbursed rather than reimbursed as part of (or incident to) the overall payment for particular services.

Contact your state Medicaid program to learn more about specific coverage in your state and see this CMS resource to learn more about Medicaid's optional and mandatory benefits.

For more information on out of state care for children, see this CMS resource.

Medicare

Medicare drug coverage for SCD treatments is described below. Note that prescription drug coverage under Part D plans varies by plan. Patients should consult their specific Medicare Part D plan for exact coverage.[43]

- **Hydroxyurea** is generally covered under Part D.[44, 45]
- **Endari (L-glutamine glutamine oral powder)** is covered under some Part D plans but may require prior authorization, have dispensing limits, and be available at limited pharmacies.[44, 45]
- **Adakveo (crizanlizumab-tmca)** is administered via IV infusion in a physician's office or outpatient setting, therefore it is generally covered under Part B. Under Part B, Medicare covers Adakveo without prior authorization when it is considered medically "reasonable and necessary."[46]

In the fiscal year (FY) 2025 Medicare Hospital Inpatient Prospective Payment System (IPPS), CMS finalized a proposal to better promote access to potentially lifesaving therapies for SCD. Specifically, CMS finalized the proposal to increase the new technology add-on payment for certain gene therapies approved for new technology add-on payments when indicated and used specifically for the treatment of SCD.

Payment and Coverage for Pain Management

Medicare

In the Calendar Year (CY) 2023 Medicare Physician Fee Schedule Final Rule, CMS created HCPCS codes describing monthly chronic pain management and treatment services.[47] These services can help Medicare beneficiaries with SCD and chronic pain to receive holistic chronic pain care within a trusting, supportive, and ongoing care relationship.

Codes for Pain Management

- HCPCS G3002: Chronic Pain Management Services
- HCPCS G3003: Add-on Code for Chronic Pain Management Services

For more information about using these codes, see:

- MLN Booklet: Evaluation and Management Services Guide (CMS)
- MLN Booklet: Chronic Care Management Services (CMS)

Payment and Coverage for Gene Therapies

Medicaid

The Cell and Gene Therapy Access Model

On January 30, 2024, the CMS Innovation Center announced the Cell and Gene Therapy (CGT) Access Model. The CGT Access Model aims to improve the lives of people with Medicaid living with rare and severe diseases by increasing access to potentially transformative treatments.[48] Initially, the Model will focus on gene therapies for SCD. The CGT Access Model will support outcomes-based agreements between states and drug manufacturers that will provide treatments within a framework that seeks to lower costs for participating states and ties payment to outcomes.

Figure 2. This figure shows the goals of the CMMI CGT Access Model

The goal of this framework is to reduce the burden of negotiating and implementing outcomes-based agreements on states, thereby facilitating savings and stability, while increasing equitable access to innovative treatment.

By doing so, this model can potentially help address the historic disparities, poor health outcomes, and low life expectancy associated with sickle cell disease. Other conditions might be added to the model over time.

To learn more about the CGT Access Model, see:

- Cell and Gene Therapy (CGT) Access Model webpage

Medicare

Allogeneic Hematopoietic Stem Cell Transplantation (HSCT) is covered under the Coverage with Evidence paradigm, in which Medicare grants coverage for certain services administered in association with approved clinical studies or clinical data collection. Under this national coverage determination, only beneficiaries with severe, symptomatic sickle cell disease who are participating in an approved prospective clinical trial are covered.[49]

Payment for Care Coordination and Promotion of Whole-Person Care

Medicaid

For Medicaid enrollees, some states provide payment for Medicaid-covered services provided by CHWs, case managers, and peer support specialists who can support people with SCD in accessing services to help identify and meet individual's health-related social needs. Contact your state Medicaid program to find out if the state covers services furnished by these providers.

Medicare

Medicare Part B provides payment for community health integration and principal illness navigation services provided by CHWs, care navigators, and peer support specialists.[50] Medicare pays for services involving CHWs to address health-related social needs that impact care. The goal is to increase access to culturally-sensitive care from CHWs, care navigators, and peer support specialists.

Medicare has also established payment for services involving caregiving training. There are five CPT® codes describing situations in which practitioners train and involve caregivers in helping to carry out treatment plans for certain Medicare beneficiaries (CPT® codes 96202, 96203, 97550, 97551, and 97552).

To learn more about CPT codes for screening, caregiver training services, and health equity-related policies in the 2024 Physician Fee Schedule Final Rule, see:

- Medicare Learning Network: Health Equity Services In The 2024 Physician Fee Schedule Final Rule (CMS)
- CY 2024 Physician Fee Schedule (CMS)

Medicare Advantage plans may offer primarily health-related complementary therapies as a supplemental benefit and non-primarily health-related complementary therapies as a Special Supplemental Benefit for the Chronically Ill (SSBCI). Complementary therapies in Medicare are those that are offered alongside what is considered traditional medical treatment.[51, 52]

Payment and Coverage for Health-Related Social Needs and Social Determinants of Health

Screening

Medicare

As of 2024, the Medicare Physician Fee Schedule includes a new stand-alone G code, G0136, to pay for administering an SDOH risk assessment, no more than once every 6 months.

Transportation

Medicaid

Access to reliable transportation is a critical factor that impacts the ability of an individual with SCD to attend health care appointments, receive treatment, and access other health care services, especially those located in rural areas.

State Medicaid plans are required to ensure that eligible Medicaid beneficiaries have necessary transportation to and from providers.[53] The Consolidated Appropriations Act of 2021 adds the assurance of transportation to Medicaid statute and limits Federal Financial Participation (FFP) for non-emergency medical transportation (NEMT) expenditures unless the state plan provides for methods and procedures that ensure payment for such services is consistent with efficiency, economy, and quality of care. Medicaid agencies are also required to offer and provide recipients of Early and Periodic Screening, Diagnostic and Treatment (EPSDT) services with necessary assistance with transportation. States may pay for transportation services as an administrative expense or as an optional service. Many states combine the two methods in order to provide maximum access.

To learn more about Medicaid's transportation supports, see:

- Assurance of Transportation: A Medicaid Transportation Coverage Guide for States (CMS)

Medicare Advantage

Medicare Advantage plans may provide transportation to non-emergent, covered Part A and Part B services as a supplemental benefit. The transportation offered must be used exclusively to accommodate the enrollee's health care needs and must be arranged, or directly provided, by the Medicare Advantage plan.[54]

Nutrition

Nutrition plays a key role in optimizing health, growth development, and immune function, and preventing acute pain episodes among individuals with SCD. It's important for individuals with SCD to eat a well-balanced diet rich in leafy greens, red and yellow vegetables, adequate fluids, and avoid refined foods that include sugars, fried food, and alcohol. Some studies also suggest that there may be benefits in using vitamin C and E supplements as well as omega 3 fatty acids.[55]

Medicaid

Through Medicaid demonstrations under section 1115 of the Social Security Act (Medicaid 1115 demonstrations), some states cover evidence-based services to address health-related social needs (HRSN) for medically appropriate populations based on clinical and social risk factors. Examples of individuals that could be served across Medicaid authorities include children and pregnant individuals identified as high risk, individuals who are or are at risk of becoming homeless, individuals with serious mental illness and/or substance use disorder, and individuals experiencing high-risk care transitions. Nutrition supports covered under certain states' demonstrations vary by state and eligibility criteria, but can include:[56]

- Case management services for access to food/nutrition;
- Nutrition counseling and instruction, which can be tailored to the needs of those with SCD;
- Home delivered meals or pantry stocking for those with certain nutrition-sensitive health conditions;
- Nutrition prescriptions, tailored to health risk, such as healthy food vouchers or fruit and vegetable prescriptions; and
- Grocery provisions for high-risk individuals to avoid unnecessary acute care admission or institutionalization

To learn more about what HRSN supports states may cover, see:

- Coverage of HRSN Services in Medicaid and CHIP Guide (CMS)
- Coverage of Services and Supports to Address Health-Related Social Needs in Medicaid and the Children's Health Insurance Program (CMS)

Medicare Advantage

Medicare Advantage plans may offer general nutritional education through classes and/or individual counseling as a supplemental benefit. In addition, Medicare Advantage plans may offer the following nutrition supports as SSBCI:[57]

- Meals provided through home delivery and/or offered in a congregate setting
- Food and produce may be covered to assist chronically ill enrollees in meeting nutritional needs

Housing and Living Conditions

Housing instability or inadequate living conditions can exacerbate the challenges associated with managing SCD, such as increased stress and compromised immune function. Environmental irritants and toxins such as mold and dust can irritate the lungs and trigger asthma attacks, increasing the risk of or worsening Acute Chest Syndrome, commonly experienced by individuals with SCD. Access to safe and affordable housing promotes better health outcomes by providing a supportive environment suitable for disease management and overall well-being for individuals with SCD.

Medicaid

Through Medicaid 1115 demonstrations, some states provide Medicaid enrollees with specific clinical and social risk factors with medically-appropriate, evidence-based, and time-limited housing supports. Examples of individuals that could be served across Medicaid authorities include children and pregnant individuals identified as high risk, individuals who are or are at risk of becoming homeless, individuals with serious mental illness and/or substance use disorder, and individuals experiencing high-risk care transitions. The housing supports covered under certain states' 1115 demonstrations vary by state and eligibility criteria but can include:[54]

- Assistance finding and securing housing, pre-tenancy navigations service, and one-time transition and moving costs;
- Short-term pre-procedure and/or post-hospitalization housing, if clinically appropriate, for up to 6 months per year, only where integrated, clinically oriented recuperative or rehabilitative services and supports are provided;
- Caregiver respite, with or without room and board;
- Assistance paying for utilities like water and electricity; for up to 6 months for medically complex individuals who meet criteria for housing supports;
- Home remediations that are medically necessary (e.g., air filtration, mold, and pest removal); and
- Home/environmental accessibility modifications (e.g., wheelchair accessibility ramps).

Medicare Advantage

Medicare Advantage plans may offer housing and living condition supports as SSBCI, such as:[57]

- Pest control
- Indoor air quality equipment and services
- Structural home modifications
- General supports for living, including:
  - Plan-sponsored housing consultations and/or subsidies for rent or assisted living communities
  - Utility subsidies

CMS's Coverage to Care program is designed to assist CMS enrollees in understanding their health coverage and connecting to primary care and preventive services.

Discuss and share the following resource with individuals with SCD:

- Roadmap to Better Care explains what health coverage is and how individuals can utilize primary care to assist them in navigating a path to better health.
- My Health Coverage at-a-Glance is meant to be customized with information specific to an individual's health plan all in one location.
- 5 Ways to Make the Most of Your Health Coverage is a quick checklist on how individuals can make the most of their health coverage.

To learn more about chronic care management, see:

- Chronic Care Management Toolkit provides resources you can use to raise awareness about the importance of chronic care management services for Medicare and dual eligible patients managing multiple chronic conditions.
- Chronic Care Management Providers Checklist includes more about chronic care management and its benefits.

5. Supporting Multi-Disciplinary Care Across Settings

Care Coordination and SCD Management in Primary Care

Care coordination and management among primary care providers (PCPs) is vital for providing comprehensive management of SCD among patients. Patients with SCD often require regular monitoring and specialized care to address both acute and chronic aspects of the disease.

Coordinated care involves PCPs and other health care professionals working together to:

- Organize patient care activities
- Implement personalized care plans tailored to each patient's unique needs
- Share information with all participants involved in caring for the patient with SCD, including sharing electronic health information with providers for care coordination and social and support service referrals.

This approach helps prevent fragmented care, reduces the risk of complications, and has the potential to improve overall health outcomes for individuals with SCD.[58]

For more information about care management, see:

- CMS Care Management, which includes FAQs about services to help address health-related social needs in the 2024 Medicare Physician Fee Schedule final rule,
- Advance Care Planning Services fact sheet
- Behavioral Health Integration Services MLN booklet
- Chronic Care Management Services MLN booklet, and
- Transitional Care Management Services MLN booklet.

Urgent and Emergency Care

People with SCD visit the emergency department an average of three times per year, often due to an acute pain episode arising from a vaso-occlusive episode.[33, 59] Share this resource with patients to support their visits to the emergency department:

- Do You Use the Emergency Department for Care of Sickle Cell Disease? What to Know Before You Go. (CDC)

Behavioral Health Care

SCD pain can have many negative impacts on patients and families, such as social isolation, finance issues, low self-esteem, and academic challenges.[55] About 1 in 3 individuals with SCD experience depression, which has been strongly associated with worse physical and mental quality of life outcomes. In addition, health care costs for these individuals were more than double those of patients with SCD without depression.[60]

Licensed behavioral health professionals should support the mental and substance use disorder needs of people with SCD by providing appropriate screening (e.g., for depression and anxiety), diagnosis, referral, and management of psychiatric disorders.[61] Primary care practices with integrated behavioral health may also wish to consult AHRQ's Integration Academy for additional resources on behavioral health integration. Primary care practices may also want to consult SAMHSA's Office of Behavioral Health Equity for additional information and technical assistance on how to reduce behavioral health disparities for their patients with SCD.

Discuss and share the following mental health support resources with individuals with SCD:

- 988 Suicide and Crisis Lifeline offers 24/7 judgment-free support for mental health, substance use, and more.
- Mental Health and Sickle Cell Disease Gene Therapy Participation (NIH) offers information about gene therapy clinical trials and how to find mental health support.

For more information about Medicare- and Medicaid-covered behavioral health services and who is eligible to provide them, visit:

- Medicare Learning Network (MLN): Medicare and Mental Health (CMS)
- Medicaid and Behavioral Health Services (CMS)

Multidisciplinary Care Teams

Children and adults with SCD often require complex multidisciplinary care and attention and multiple care providers to provide adequate treatment for their needs.[62]

SCD health care providers range from primary care physicians to hematologists along with other members of the care team. Ophthalmologists, dentists, and obstetricians/gynecologists are typically only required to have some "knowledge of evidence-based management of individuals with SCD," with each specialty having different requirements regarding their screening and treatment practices for SCD related complications.[61]

Multidisciplinary teams can address the diverse needs of patients with SCD and provide more effective care by using a **shared decision-making approach**, which involves:[63]

- A collaborative effort between patients with SCD, families, and health care professionals;
- Selecting the most appropriate treatment or management plan tailored to the patient's preferences and values;
- Supporting the individual's autonomy;
- Respecting the individual's competence and interdependence on others; and
- Establishing trust in the relationship.

For additional resources on using a shared decision making approach, please visit AHRQ's Shared Decision Making website.

CHWs, Patient Navigators, and Peer Specialists

Non-clinical health professionals play a crucial role supporting individuals with SCD by assisting them in navigating the complexities of the health care system and providing different types of support. For example:

- **CHWs**, commonly known as outreach workers, typically are lay members of the community that work in community settings. They often share the same ethnicity, language, and life experiences as an individual with SCD and provide culturally appropriate education, interpretation and translation services, informal counseling, and help individuals with SCD advocate for their health needs.[64]
  - HRSA funds the Sickle Cell Disease Newborn Screening Follow-up Program with community-based organizations (CBOs) that support individuals with SCD and their families and link them to community resources and evidence-based care, strengthening the linkage from community to specialty care. Training is provided to CHWs on specific SCD curriculum tailored to the needs of the population. By facilitating access to quality SCD care, the program aims to improve the care and reduce the rates of loss to follow-up of individuals identified with SCD.
- **Patient navigators**, Patient navigators, often called care coordinators or health navigators, typically work within a hospital or clinic to reduce barriers to health care among individuals with SCD and promote continuity of care. They help facilitate appointments, coordinate referrals, and ensure access to essential services (including culturally/linguistically appropriate services), social determinants of health, and other support programs.[65]
- **Peer specialists** are individuals with lived experience related to SCD that can offer empathy, understanding, and practical advice to individuals with SCD, their caregivers, and families.

6. Resources for Individuals with Sickle Cell Disease and Those Who Support Them

CMS has compiled a list of resources to help individuals with SCD), the health care professionals who treat them, their families, caregivers, and supporting community-based organizations successfully understand their health coverage, navigate care, and access supports.

General Information About SCD

SCD is a group of inherited genetic blood cell disorders. Individuals with SCD can experience a variety of health challenges such as severe acute pain episodes, chronic pain, infections, and organ failure. For general resources about SCD, see:

- What is Sickle Cell Disease? (NIH) introduces the disease and links to a SCD fact sheet.
- What You Should Know About Sickle Cell Disease (CDC) includes an overview and the causes of SCD, health complications associated with SCD, and treatment options.
- About Sickle Cell Disease (CDC) provides and overview of the disease and a list of key organizations of interest to people living with SCD and their families.

- Stories of Sickle Cell (CDC) allows you to learn about the unique stories and diverse identities of individuals with SCD through short stories.
- Sickle Cell Disease Programs (HRSA) provides a list of funded community based organizations across the United States.

Understanding Health Coverage

The majority of individuals with SCD are enrolled in Medicaid/CHIP, Medicare, or insurance plans purchased through the Marketplace.[24, 66, 67] CMS's Coverage to Care program is designed to assist individuals to understand their health coverage and connecting to primary care and preventive services. For additional Coverage to Care resources, see:

- Roadmap to Better Care explains what health coverage is and how patients can utilize primary care to assist in navigating a path to better health.
- My Health Coverage at-a-Glance is meant to be customized with information specific to an individual's health plan in one location.
- 5 Ways to Make the Most of Your Health Coverage is a quick checklist on how individuals can make the most of their health coverage.

For more information on enrollment and coverage for Medicaid/CHIP, Medicare and the Marketplace, see:

- Your Medicaid or CHIP Coverage page explains how to prepare for the renewal process and includes a map to select your state to get Medicaid enrollment contact information.
- The Medicare.gov page has information on how to get started with Medicare, find health or drug plans, find care providers or contact someone with questions about your coverage.
- Healthcare.gov provides information on the Health Insurance Marketplace plans and savings if you no longer quality for Medicaid or CHIP or have certain life events or income.
- State Health Insurance Assistance Program counselors provide help to Medicare beneficiaries, their families, and caregivers.

Health and Wellness for People with SCD

The following resources are designed to support achieving holistic well-being, from disease management tools for general healthy living to mental health support services.

General Healthy Living:

- Healthy Living with Sickle Cell Disease (CDC) includes guides on how to support school-aged children and college students with SCD as well as tips for how you can prevent infection and knowing when to seek emergency care.
- Steps to Better Health Toolkit CDC) is a series of easy-to-read fact sheets for people with SCD and increase awareness about the steps to better health for people with SCD.

- Living With Sickle Cell Disease (NIH) provides steps for how to help relieve symptoms of SCD and manage the condition at home.
- Athletes: Don't Get Sidelined by Sickle Cell Trait! Play it Safe with These Helpful Tips! (CDC) addresses commonly asked questions about sickle cell trait, participation in sports, exercise related illness, and what to do to stay safe and healthy while engaging in physical activity.

Mental Health and Wellness:

Living with a chronic disease can negatively impact the mental health and wellbeing of those living with SCD, their families, and caregivers. To find support, see:

- Mental Health and Sickle Cell Disease Gene Therapy Participation (NIH) offers information about gene therapy clinical trials and how to find mental health support.
- 988 Suicide and Crisis Lifeline offers 24/7 judgment-free support for mental health, substance use, and more.

SCD Treatments

Common treatments for SCD often aim to prevent or minimize complications and acute pain episodes associated with the disease. In addition, newly approved cell-based gene therapies are one-time treatments which the potential to transform the lives of people with SCD. To learn more about treatments for SCD, see:

- The Sickle Cell Disease Treatment (NIH) webpage reviews common treatments for SCD and associated complications.
- Hydroxyurea Use for Sickle Cell Disease Fact Sheet (NIH) provides facts about hydroxyurea, potential benefits and risks, and questions to ask your health care provider.
- Understanding Gene Therapy Approaches (NIH) describes the components of gene therapy, how these components differ and other treatment options for SCD.

Pain Management

The most common complication of SCD is pain (both acute and chronic), and acute pain episodes are a leading reason for visits to the emergency department among people with SCD. To learn more about managing pain and strategies for discussing pain management with your health care provider, see:

- Steps to Better Health Toolkit: Managing Chronic Pain (CDC) reviews options for treating chronic pain and discusses the risks and benefits of opioid therapy.
- Steps to Better Health Toolkit: Managing Acute Pain (CDC) reviews options for treating acute pain and gives guidance on how to work with your health care provider to create an individualized pain management plan.
- Managing Pain with Sickle Cell Disease Fact Sheet (NIH) provides tips to manages your pain and prevent serious problems.
- CMS Sickle Cell Disease Action Plan (CMS) addresses challenges specific to SCD and lists programs aiming to improve health outcomes and reduce health disparities for individuals with SCD.

Urgent and Emergency Care

Individuals aged 17-59 with SCD visit the emergency department an average of three times per year, often due to an acute pain episode. For tips on receiving better care in the emergency department and when leaving the hospital, see:

- Do You Use the Emergency Department for Care of Sickle Cell Disease? What to Know Before You Go. (CDC)
- Taking Care of Myself: A Guide for When I Leave the Hospital (AHRQ)

Telehealth

- Telehealth.HHS.gov providers resources for patients to understand what telehealth is and how to access it. Telehealth may be particularly beneficial for individuals in rural areas or who do not have access to a nearby specialist.

Maternal and Reproductive Health

SCD can have a significant impact on a woman's reproductive health. During pregnancy, women with SCD are at higher risk for developing complications, such as preeclampsia (high blood pressure during pregnancy) and blood clots, than women without SCD. For some women with SCD, pregnancy can make their disease more severe, and treatments may need to be changed to help manage their complications.[68]

The following resources provide guidance for individuals on maternal health and during all phases of pregnancy, including preconception, prenatal, and postpartum care:

- Pregnancy, Reproduction, and Sickle Cell Disease (NIH) provides information on the ways in which SCD can impact reproduction and pregnancy.
- Women with Sickle Cell Disease and Preconception Care: What to Know Before Getting Pregnant (CDC) offers information if one is thinking about or planning to get pregnant, education around preconception care, and steps for staying healthy during a future pregnancy.
- Women with Sickle Cell Disease and Prenatal Care (CDC) focuses on the steps you can take during your pregnancy to stay healthy.
- Women with Sickle Cell Disease and Postpartum Care: What to Know After Delivering Your Baby (CDC) describes steps that can be taken to support physical, emotional, and mental health as you recover and bond with your baby.
- Fact Sheet on In Vitro Fertilization Use Across the United States (ASPE) provides information on assisted reproductive technology options that may help individuals with SCD conceive or support fertility preservation.
- CDC Sickle Cell and Pregnancy page includes what you need to know about how SCD affects pregnancy.
- NIH Pregnancy, Reproduction and Sickle Cell Disease page provides information on family planning with SCD.

Conclusion

Health care providers, as well as community partners, play a key role in providing care, improving care management, and supporting the varied needs of individuals with SCD. This toolkit is designed to assist the care team in supporting individuals with SCD by introducing SCD, including the common barriers to care, and summarizing CMS program coverage for services and new SCD treatments. CMS programs help mitigate health disparities and improve the overall well-being of individuals with SCD.

As health care providers continue to engage with individuals affected by SCD, it is essential to stay up to date on emerging research, advancements in treatment modalities, and evolving standards of care. By staying informed and adaptable, health care providers can ensure that their practices reflect the latest evidence-based guidelines and promote the best possible outcomes for patients with SCD.

This toolkit represents a step forward in advancing the quality and accessibility of care for individuals with SCD. Health care providers and other members of the care team are encouraged to share the educational resources in this toolkit with the individuals they serve and with other providers who may benefit.

Appendix: The Basics of Sickle Cell Disease (SCD)

For general information about SCD, see:

- What is Sickle Cell Disease? (NIH)
- About Sickle Cell Disease (CDC)
- What You Should Know About Sickle Cell Disease (CDC)
- Sickle Cell Disease: What You Need to Know (CMS)
- Sickle Cell Information for Healthcare Providers (CDC)
- Sickle Cell Disease Programs (HRSA)

What is SCD?

SCD is a group of inherited genetic blood cell disorders. Individuals with SCD can experience a variety of health challenges such as severe pain episodes, infections, and organ failure.[69]

Figure 1. Prevalence of Sickle Cell Disease by State[70]

Geographic distribution of SCD by state using data derived from the National Newborn Screening Information System.

Epidemiology of SCD

SCD affects over 100,000 Americans in the United States and disproportionately impacts Black or African American individuals and Hispanic or Latino individuals (Centers for Disease Control and Prevention, 2024a). It can also be found among individuals of African, Southern European, Middle Eastern, or Asian Indian ancestry.[71] The disease is unevenly distributed across the U.S., with Southern and Mid-Atlantic states having the highest rates of the disease. Medicaid is the predominant source of health coverage for people with SCD.[72] Individuals with SCD are more likely to be dually eligible for Medicare and Medicaid than those without SCD.[55, 61]

For more information on SCD prevalence, see these CMS Data Highlights:

- The Invisible Crisis: Understanding Pain Management in Medicare Beneficiaries with Sickle Cell Disease
- Prevalence of Sickle Cell Disease among Medicare Fee-for-Service Beneficiaries, Age 18-75 Years, in 2016
- Prevalence of Sickle Cell Disease among Medicaid Beneficiaries in 2012
- At a Glance: Medicaid and CHIP Beneficiaries with Sickle Cell Disease (SCD), T-MSIS Analytic Files (TAF) 2017

SCD Causes and Risk Factors

SCD begins at birth when a child receives two mutated hemoglobin genes (one from each parent) that code for abnormal hemoglobin. A mutation in the HBB gene causes SCD. This mutation leads to production of abnormal hemoglobin, known as Hemoglobin S (HbS). When oxygen levels are low, HbS molecules can cause red blood cells to become rigid in a "sickle" like shape).[69] If an individual inherits sickle cell gene from one parent and a normal gene from the other parent, it indicates they have sickle cell trait, which more than 2 million in the U.S. live with.[73] These individuals are at greater risk for passing the gene to their children.

Common Types of SCD[1]	Rare Types of SCD[48]
• HbSS • HbSC • HbS beta thalassemia	• HbSD • HbSE • HbSO

For more information on SCD causes and risk factors, see this NIH resource:

- Sickle Cell Disease Causes and Risk Factors (NIH)

100k+ People affected in the U.S.

Sickle cell trait appears in 1 in 13 Black or African American births

Disease occurs in 1 in 365 Black or African American births

Disease occurs in 1 in 16,300 Hispanic or Latino births

(Centers for Disease Control and Prevention, 2024a)

Symptoms

Individuals with SCD typically begin developing symptoms within their first year of life (usually around 5-6 months of age). Common early symptoms of SCD include jaundice of the skin or eyes, swelling in the hands and feet, and extreme tiredness or fussiness.[74] SCD is a lifelong illness.[1,71] Currently there is no way to prevent SCD because it is genetic condition. Advances in the medical field including newborn screening, penicillin prophylaxis, and other therapeutics have shifted SCD from a fatal childhood disease to a chronic condition with long term health complications (e.g., acute chest syndrome, stroke, and organ damage).[75, 76, 77] However, individuals with SCD still face higher rates of morbidity and mortality compared to those without the condition.[78]

The presentation and severity of SCD symptoms can vary from person to person, change over time, and adversely affect an individual's physical and mental health. Individuals with SCD often have multiple chronic conditions which require access to therapies that prevent and treat SCD complications, minimize hospitalizations, and lower symptom burden.

Symptoms and complications of SCD can affect the whole body and may include:[79]

- Acute Chest Syndrome
- Anemia
- Avascular Necrosis
- Behavioral health
- Blood Clots
- Dactylitis
- Fever
- Infection
- Kidney Problems
- Leg Ulcers
- Liver Problems
- Organ Damage
- Pain
- Priapism
- Pulmonary Hypertension
- Sleep-Disordered Breathing
- Splenic Sequestration
- Stroke
- Vision Loss

For more information on SCD symptoms and complications, see these resources:

- Sickle Cell Disease Symptoms (NIH)
- Complications of Sickle Cell Disease (CDC)

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Paid for by the U.S. Department of Health and Human Services
August 2024

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