

A Public Health Strategic Plan to Address Sickle Cell Disease Across the Lifespan

2015—2018

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Executive Summary

Sickle cell disease (SCD) is an inherited disorder of hemoglobin (Hgb). It is one of the most prevalent genetic disorders detected in Michigan and has a dramatic impact on the lives of over 2,800 residents with the disease and their families. The health outcomes and disparities resulting from SCD in Michigan make SCD a public health priority for the Michigan Department of Health and Human Services (MDHHS). The MDHHS public health strategic plan to address SCD is a landmark accomplishment in many ways for individuals (both children and adults) and families affected by a sickling condition. It aims to focus attention on both the MDHHS Newborn Screening Program (NBS) and Hemoglobinopathy Quality Improvement Program (HQIP) with a sense of need and urgency for this vulnerable population.

This plan recommends a framework based on the findings of a Needs Assessment conducted from June through October 2014. A joint departmental effort with the Lifecourse Epidemiology & Genomics Division (LEGD), the Family & Community Health Division, and Children's Special Health Care Services (CSHCS) provided the impetus and staff needed to accomplish this strategic planning project. This plan provides a set of public health interventions to reduce the burden of SCD in the state through improved awareness, comprehensive transitional care programs, and increased utilization of community mental health and behavioral health services.

Seven key gaps were identified as public health priorities by planning participants. These included: 1) a need for education to emergency department (ED) physicians and ED guidelines to improve acute care for patients; 2) a lack of comprehensive transitional care programs; 3) underutilization of community mental health and behavioral health services for psychosocial needs; 4) lack of use and adherence to disease-modifying therapies such as hydroxyurea; 5) a need for better coordination of SCD research efforts; 6) more statewide-level education to increase awareness of SCD, sickle cell trait (SCT), and medical therapies; and 7) a lack of providers who treat all aspects of SCD, with an emphasis on increasing adult providers.

Recurring themes were extracted from the key gaps and reduced to seven goals, eight strategies, and detailed tactics that will be used to accomplish each goal. The goals promote integration and expansion of SCD services and resources within existing public health programs, and emphasize the need to enhance patients' clinical experience. *The goals are summarized below:*

- ◆ **Education & Awareness:** Develop a statewide multi-level messaging and communication strategy to increase awareness of SCD, disease-modifying medical therapies, and SCT.
- ◆ **Transition:** Develop and implement strategies for improving transition from pediatric to adult care.
- ◆ **Provider Shortage:** Increase availability of primary and specialty care providers who treat all aspects of SCD with an emphasis on increasing providers for adults.
- ◆ **Psychosocial/Mental Health Support:** Increase recognition of the need to address psychological issues and provide mental health support.
- ◆ **Medication Adherence:** Develop protocols for improving medication adherence in accordance with the National Heart, Lung and Blood Institute's (NHLBI) Evidence-Based Management of SCD Guidelines.
- ◆ **Day Treatment Clinics:** Improve acute care in the emergency room or alternative settings.
- ◆ **Research:** Establish a Michigan SCD Consortium to coordinate public health research efforts within the State of Michigan.

Implementation will leverage upon existing opportunities and initiatives that complement and coordinate with the mission and vision of MDHHS.



Acknowledgements

The state public health SCD strategic plan is the result of many months of collaborative efforts by nearly 100 individuals. A special acknowledgement and heartfelt thank you is extended to everyone - focus group participants, hematologists, pediatricians, workgroup members, advisory committee members, reviewers, staff, individuals living with SCD and their parents, who gave their time and insight to shape this consensus plan. Contributors to the strategic planning meeting included the following individuals:

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Acronyms

Acute Chest Syndrome	ACS
Child Adolescent Health Center	CAHC
Children Special Health Care Services	CSHCS
Children’s Hospital of Michigan	CHM
Detroit Medical Center	DMC
Division of Immunizations	DOI
Emergency Department	ED
Family Center for Children & Youth With Special Health Care Needs	FCCYSHCN
Family Medical Leave Act	FMLA
Health Status Assessments	HSA
Hemoglobin	Hgb
Hematology Quality Improvement Committee	HemQIC
Hemoglobinopathy Quality Improvement Program	HQIP
Lifecourse Epidemiology & Genomics Division	LEGD
Maternal Child Health	MCH
Michigan Department of Health & Human Services	MDHHS
Newborn Screening	NBS
Red Blood Cells	RBC
Registry & Surveillance System for Hemoglobinopathies	RuSH
Sickle Cell Disease	SCD
Sickle Cell Disease Association of America - Michigan Chapter	SCDAA-MI
Sickle Cell Trait	SCT
University of Michigan	UM
Women, Infant, and Children’s Supplemental Nutrition Program	WIC



Introduction: Development of the Sickle Cell Strategic Plan

As part of the MDHHS Population Health & Community Services Administration strategic planning process during 2013, a need to address SCD was identified which led to development of **Strategic Priority 2.4: Design a public health approach to SCD across the lifespan**. Statewide, approximately 2,800 Michigan residents live with SCD and approximately 140,000 people carry the SCT.

Beginning June 2014, the MDHHS SCD Strategy Team engaged in a mixture of planning activities in collaboration with the Sickle Cell Disease Association of American - Michigan Chapter (SCDAA-MI), the Michigan Hematology Quality Improvement Committee (HemQIC), hematologists/nurses/ social workers, and individuals directly impacted by SCD. MDHHS staff throughout the department including Immunization, Medicaid, Maternal & Child Health (MCH), the Women, Infant, and Children's Supplemental Nutrition Program (WIC), Children's Special Health Care Services (CSHCS) and the Laboratory also participated in the planning process.

The planning process included the five steps described below. The process was designed to gather pertinent findings and experiences from research literature, community members, individuals with sickle cell disease, and healthcare providers.

- 1) Relevant literature and other state strategic plans were reviewed.
- 2) The HemQIC was convened to gather input on important issues impacting the SCD population, current and needed clinical medical management resources, underutilized opportunities to improve quality of life, critical public health focus areas, and health insurance opportunities to improve quality and access.
- 3) Five focus group meetings were conducted in Detroit, Saginaw, Lansing, Grand Rapids, and Benton Harbor with individuals living with SCD and/or their caregivers. Recurring themes were extracted by the Strategy Team and reduced to seven focus areas - day treatment, transition, psychosocial/mental health, medication adherence, research, public education/awareness, and public/private insurance challenges.
- 4) The focus areas were used to identify workgroup topics for discussion during the stakeholder strategic planning meeting held in October 2014.
- 5) Workgroups were formed for the stakeholder meeting and participants identified critical issues, actions to include in the plan, and developed recommendations.

Information collected through the planning process was summarized by the Strategy Team into this strategic plan, accompanied by recommended action steps. Subsequently, the full plan was submitted in draft form to MDHHS partners for review and further comment. Over the course of three years, beginning in the fall of 2015, the department will begin implementing parts of the plan to improve the care of Michigan residents living with SCD.



Key Partners for Achieving State Plan Goals

The MDHHS Lifecourse Epidemiology & Genomics Division will continue our long-term collaboration with internal and external partners to improve the health of individuals with SCD. Benefits from the partnerships below will include leveraging and maximizing resources, utilizing existing expertise, and increasing capacity.

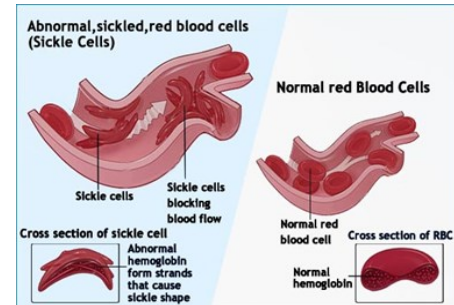
- ◆ **Child & Adolescent Health Centers (CAHC)** : A program jointly-funded by the MDHHS and Michigan Department of Education (MDE) to promote the health of children, adolescents and their families by providing primary and preventative care, comprehensive health assessment, vision and hearing screening, medication, immunizations, treatment of acute illness, co-management of chronic illness, health education and mental health care. There are currently 72 school-based, 10 school-linked, as well as 17 school-wellness centers in Michigan.
- ◆ **Children’s Special Health Care Services (CSHCS)**: A program within the MDHHS for children and some adults with special health care needs and their families. CSHCS helps persons with chronic health problems by providing coverage and referral for specialty services, family centered services to support primary caretakers, community-based services to help caregivers care for children at home, and service coordination to pull together the services of many providers who work within different agencies.
- ◆ **Family Center for Children and Youth with Special Health Care Needs (FCYSHCN)**: A program within CSHCS whose primary purpose is to help shape CSHCS policies and procedures and to help families navigate the CSHCS system. Through its *Parent to Parent Support Network*, the Center provides emotional support and information statewide to families of children with special health care needs.
- ◆ **Division of Immunization (DOI)**: A program within the MDHHS that develops strategies to ensure children and adults in Michigan are appropriately immunized and have access to vaccines. The program is responsible for a variety of wide-ranging functions, including vaccine management, quality assurance, analysis of immunization rates, identification of and response to immunization disparities, and vaccine-preventable disease surveillance and control.
- ◆ **Maternal Infant Health Program (MIHP)**: An MDHHS program that provides home visitation support and care coordination for pregnant women and infants on Medicaid. Services supplement regular prenatal/ infant care and assist health providers in managing the beneficiary’s health and wellbeing.
- ◆ **Michigan Hemoglobinopathy Quality Improvement Committee (HemQIC)**: This committee consists of pediatric and adult hematologists, nurses, NBS and laboratory staff who review the LEGD system for diagnosis and treatment services provided to newborns and children with hemoglobinopathies detected by Michigan NBS, other states or later diagnosis. This includes development of diagnostic protocols, medical management protocols, short and long-term follow-up protocols and database management.
- ◆ **Sickle Cell Disease Association of America, Michigan Chapter (SCDAA-MI)**: Through a contract with MDHHS since 1987, the SCDAA-MI has assured that newborns with sickle cell conditions identified through NBS receive comprehensive diagnostic, education, counseling, social work, laboratory testing and treatment services.
- ◆ **Specialty Clinics**: Hematology clinics in Ann Arbor, Detroit, Flint, Lansing, Kalamazoo, and Grand Rapids that provide multi-disciplinary care for sickle cell and thalassemia patients.
- ◆ **Women, Infant, and Children’s Supplemental Nutrition Program (WIC)**: MDHHS/WIC contracts with local agencies that have responsibility for providing direct program services to clients. Services provided include nutrition education, food benefits, breastfeeding support and referrals through local health departments or other community agencies.



Background Information on Sickle Cell Disease and Trait

Sickle cell disease is an inherited disorder that affects the formation of hemoglobin (Hgb), the protein in red blood cells that carries oxygen. The red blood cells (RBC) become hard, sticky and shaped like a farmer's sickle. These sickled cells block blood and oxygen flow in blood vessels, and break down more rapidly than normal RBCs. This can cause a low blood count (anemia).

SCD is more common among people whose ancestors come from sub-Saharan Africa, Spanish speaking regions in the world (South America, Cuba, and Central America), Saudi Arabia, India, and Mediterranean countries such as Turkey, Sicily, Greece, and Italy.¹ Statewide, approximately 2,800 Michigan residents live with SCD and approximately 140,000 people carry the SCT (one copy of the sickle cell gene). The NBS program identifies approximately 65 newborns with hemoglobinopathies per year.



Source: <http://microbeonline.com/relationship-sickle-cell-anemia-malaria/>

Complications of Sickle Cell Disease

SCD can block the flow of blood in arteries in many parts of the body, causing a variety of complications. The hallmark symptom of SCD is *sickle cell crisis*, which causes sudden attacks of severe pain. An infection or blockage of blood vessels in the lungs can lead to acute chest syndrome (ACS), another common and serious occurrence.² The abnormal shape of red blood cells found in patients with SCD contributes to co-morbidities throughout the lifespan including pneumococcal infections and acute spleen sequestrations in infants, pulmonary hypertension, stroke, gallbladder disease, and organ damage. SCD is also associated with premature mortality.³

Sickle Cell Trait

SCT is different from SCD. Individuals with SCT cannot develop SCD later in life; however, they can pass the sickle cell gene to their children. SCT affects 1 in 12 Blacks or African Americans in the United States and occurs when a person inherits a sickle cell gene from just one parent. SCT and SCD inheritance is as follows:⁴

- ◆ If both parents have SCT, there is a 50% (or 1 in 2) chance that any child of theirs will have SCT.
- ◆ If both parents have SCT, there is a 25% (or 1 in 4) chance that any child of theirs will have SCD. There is the same 25% (or 1 in 4) chance that the child will not have SCD or SCT.
- ◆ If one parent has SCT, there is a 50% (or 1 in 2) chance that any child of this parent will have SCT and an equal 50% chance that the child will not have SCT.⁵

In rare cases, environmental situations that lead to low oxygen levels in the air or dehydration can lead to possible complications for people with SCT.⁴



Challenges of the Disease

For many who have SCD, daily living can consist of illness, pain, time-consuming trips to the ED, stigmatization, and a compromised quality of life. This public health plan addresses challenges by responding to gaps identified by patients, affected families, advocacy organizations, health care providers, public health practitioners and others involved in the delivery of services to those with SCD or trait.

SCD affects all aspects of patients' lives. The debilitating symptoms and complex treatment needs can limit their ability to perform in school, pursue careers, have a family, and maintain relationships. The disease takes an emotional toll as patients face challenges with the healthcare system, stigma within society, financial hardships, and worry about their future. Both young and old live with constant reminders they are not able to live a normal life. Complications of SCD include daily fatigue and cognitive effects, and the long-term progressive damage of SCD.

The challenges in adequately managing SCD are much broader than just the availability of medical treatments. Research has shown that patients have difficulties having their condition recognized or their symptoms taken seriously, navigating the healthcare system, interacting with healthcare professionals, getting access to treatment, and getting needed accommodations within their schools and workplaces.

"When I was a child the doctors and nurses felt bad for me having sickle cell disease. They were nice, thought I was a cute kid. Once I turned 18, all of that stopped and I was treated totally different. That's when they started saying I was a drug addict and there was nothing they could do for me."

-Focus Group Participant

Planning participants identified the following gaps as public health priorities, all of which have been incorporated into the statewide plan:

- ◆ A need for education of ED physicians and recommended ED guidelines to improve acute care for patients.
- ◆ Lack of comprehensive transitional care programs.
- ◆ Underutilization of community mental health and behavioral health services for psychosocial needs.
- ◆ Lack of adherence and underutilization of disease-modifying therapies such as hydroxyurea.
- ◆ A need for more and better coordinated public health SCD research studies in Michigan.
- ◆ A need for more statewide-level education to increase awareness of SCD, SCT, and medical therapies.
- ◆ Lack of providers who treat all aspects of SCD, especially adult providers.



Sickle Cell Surveillance in Michigan

For detailed analysis see Sickle Cell Disease Newborn Screening Surveillance Report at: http://www.michigan.gov/documents/mdch/RuSH_Final_Draft_11.07_473558_7.pdf

MDHHS staff, the SCDA-MI, and members of the HemQIC collaborated to assess the burden of Hgb disorders on individuals of all ages identified primarily through Michigan's NBS Program. From 2009-2013, systematic data collection was a priority in Michigan as part of the Centers for Disease Control & Prevention (CDC) - funded Registry and Surveillance System for Hemoglobinopathies (RuSH) grant performance measures and health status indicators. Subsequently, key baseline measurements relevant to important aspects of SCD in the state were developed, and provided important information to identify gaps in services, representing an essential step for planning public health action.⁶

After extensive data analysis and literature review, twelve hemoglobinopathy baseline measures were identified related to: incidence, demographics, confirmatory diagnosis, antibiotic prophylaxis, patient education, immunizations, public insurance enrollment, transcranial doppler screening, hydroxyurea treatment, ED visits, inpatient hospitalizations, and outpatient visits. These data span various time periods and were obtained from birth certificate records, the SCDA-MI's records on children and adults served through the center, immunization information from the MI Care Improvement Registry (MCIR), and Medicaid and CSHCS claims from the Michigan Data Warehouse. Michigan data provided benchmarks for hemoglobinopathies related to Healthy People 2020 objectives. Further, the findings provided important information to identify gaps in services, and represent an essential step for planning public health action.

Data findings from 2013 are as follows:⁷

From 1988-2013, a total of 1,733 newborns in Michigan were detected and confirmed as having SCD through the NBS Program. In 2013, 56 newborns were diagnosed with SCD; including 34 cases of sickle cell anemia, 14 of SC disease, and 8 of sickle beta thalassemia. An additional 2,772 newborns were identified as having SCT on initial screening results.

Notable findings for children and youth with SCD include:⁶

- Of those born from 2007-2011, 73 percent had penicillin prophylaxis treatment initiated within 120 days of birth, and 90 percent had penicillin prophylaxis at some point before age five.
- The pneumococcal vaccine series had the lowest completion rate (52%) while the hepatitis B series had the highest (92%). Infection is a significant contributor to morbidity and mortality in SCD. In addition to receiving routine childhood immunizations, all SCD individuals are recommended to receive pneumococcal and hepatitis B vaccines.
- Based on Michigan Medicaid claims data, the most common users (19%) of hydroxyurea among children were 15-18 year olds.
- In 2011, approximately 20 percent of children age 0-18 years had a claim for transcranial doppler screening (TCD).
- The percent of children with four or more claims for ED visits increased from 16 percent in 2008 to 19 percent in 2011.

Based on demographics from NBS, the majority of individuals with SCD live in the cities of Detroit, Saginaw, Flint, Benton Harbor, Grand Rapids, Muskegon, Kalamazoo, Benton Harbor, Lansing, and Jackson, making them high priority areas for services to individuals with SCD and their families.⁶



Key Findings

Key Findings from the planning process can be framed in terms of seven core areas. These areas include: 1) Education & Awareness; 2) Transition; 3) Provider Shortage; 4) Psychosocial/Mental Health Support; 5) Medication Adherence; 6) Day Treatment Clinics; and 7) Research. Both quantitative and qualitative information was captured in order to identify the major needs summarized below.

◆ **EDUCATION & AWARENESS:** Public education and awareness play a key role in improving the lives of people living with SCD as well as understanding among the general public. There is a general lack of understanding of SCD, thus the disease tends to be accompanied with some degree of judgment and stigmatization. A targeted increase in education and awareness could aid in general understanding and empathy of the difficulty of living with SCD. Public health awareness campaigns such as “know your status” can play a role in spreading general information about SCD and SCT. Blood drives, information pamphlets, and increased social media involvement are all different strategies that have the potential to increase public education and improve societal awareness of SCD. In addition, school-based clinics present a unique opportunity for teachers and school nurses to educate children about SCD.

◆ **TRANSITION:** Appropriate transition of care from pediatric to adult medicine and hematology care is crucial in treating sickle cell patients and managing their symptoms. The highest rate of morbidity of SCD is between the ages of 18-30, as young adults struggle through the transition of care.⁸ For a variety of reasons, sickle cell care often becomes fragmented as young adults transition from pediatric to adult care. First, there is an overall lack of adult hematologists, which leads to patients using the ED as their primary method of care. Second, ED doctors and nurses are not as well-versed in SCD as hematologists.

“I feel like I was handed to the wolves. When my cousin turned 15, two hematologists started seeing her together. An adult and pediatric hematologist. Then the pediatric hematologist just faded out and it was a seamless transition.”

-Focus Group Participant

A multi-faceted approach must be used in order to improve the transition of care for people living with SCD. Health providers, family members, and patients need to increase the level of education given at each stage of the disease. Children must learn to advocate for themselves and manage their disease at a young age in order to navigate the health system as an adult. This requires both the parents and patients taking responsibility for their own care so they can learn to navigate the medical system. Additionally, parents should adopt a “talk early/talk often” strategy to teach their children how to navigate the medical system, educate children on insurance policies and management, and teach children the importance of addressing ongoing health needs such as genetic counseling, oral health, maternal and reproductive health.

◆ **PROVIDER SHORTAGE:** There is a clear lack of providers for either adult primary care or subspecialty care. The majority of hematologists treating SCD in the state provide care to pediatric patients. There is one major clinic in Detroit solely dedicated to management of adult sickle cell patients. However, it is unclear where the remaining adult patients receive care. Thus, young adult patients with SCD find themselves in a situation where they may choose to rely on the ED for care rather than a primary care provider. Further, reports in the literature suggest that ED staff report having limited knowledge and awareness about SCD, which could result in delays in treatment and lower quality care.⁹ Research indicates adult patients with SCD who experience such discrimination or stigma are more likely to have poor health and quality of life outcomes.⁹ Existing initiatives could be explored as ways to increase the number of physicians treating patients with SCD.



Key Findings cont.

- ◆ **PSYCHOSOCIAL/MENTAL HEALTH SUPPORT:** Psychosocial and mental health care is an integral component of treating SCD. A statement in the NHLBI publication, “The Management of Sickle Cell Disease,” clearly identifies the importance of psychosocial intervention in the holistic care of the disease.¹⁰ It says, “The pain experienced by many patients with SCD can be demoralizing and overwhelming. In addition to the psychological effects of inadequately treated pain, patients have the added stress of continually searching for effective pain relief, resulting in frequent emergency room visits and episodic care. This cycle can lead to depression, which is highest among the chronically ill and in the 20-40 age group, and is often not recognized or addressed.”

Individuals with SCD can experience complications such as hypoxia, infections, delayed growth and puberty, and stroke beginning in infancy and throughout life that can lead to cognitive deficits and academic deficits. In addition, children with SCD have more school absences due to recurrent provider visits, pain crises and socioeconomic factors. Combined, these factors can lead to reduced academic attainment, as measured by grade promotion/retention rates and rates of placement in special education services. One study found that a significantly higher proportion of children with SCD were held back a grade and received special education services relative to national, state, and local school district norms for African American students.¹¹ By promoting mental health services and education, professionals treating these individuals may ensure SCD patients receive a holistic approach to care.

- ◆ **MEDICATION ADHERENCE:** SCD is associated with substantial morbidity, premature mortality, individual suffering, healthcare costs and loss of productivity.¹² There is no universal cure for SCD and treatment options are rather limited. Antibiotic prophylaxis is used to prevent infections, especially in children.¹³ Other therapies aim to minimize the effects of disease symptoms. Pain crises are managed primarily with analgesia, and hydration.¹³ Blood transfusions may be required for stroke and other complications. Bone marrow transplantation is a possible cure, however among other criteria, this requires a matched donor, and unfortunately is not feasible for many affected individuals.¹⁴

“My son was going to the hospital frequently. His doctor said that he was a good candidate for hydroxyurea. He had some adverse effects early. Now his pain crises are far less frequent. He still gets some headaches, but we’re working on it.”

-Focus Group Participant

Hydroxyurea has been found to be very effective in reducing the ‘sickling’ process and consequently the frequency of pain and hospitalizations experienced by patients.¹⁵ It is the only disease-modifying therapy for SCD and is efficacious in reducing complications such as ACS and improving survival. It is however, vastly underutilized and poorly adhered to because of individual barriers at the provider, treatment, socioeconomic, and patient levels. There is an overall lack of uniform provider practice, mistrust in treatment options, and conflicting provider messages about hydroxyurea therapy. Interventions will focus on patient/provider education and strategies to improve adherence.

- ◆ **DAY TREATMENT CLINICS:** Patients living with SCD often suffer acute pain crises and seek help in a hospital ED. This approach to pain management has proven costly and there are often serious delays prior to initiating treatment; In addition, unnecessary diagnostic procedures may be ordered. As an alternative to treatment in the ED, the sickle cell hospital day treatment clinic is often available during non-traditional clinic hours, and may be a more cost-effective approach for prompt, aggressive and specific treatment of acute pain crises. *(Continue on page 14)*



Key Findings cont.

- ◆ **DAY TREATMENT CLINICS Continued:** These clinics, when established, have been demonstrated to: (1) provide better pain management with treatment delivered, on average, in less than half the time of an ED visit; (2) reduce the number of ED visits; (3) reduce the need for hospital admission and length of stay when treated by SCD day clinic staff (which resulted in a savings of ~\$1.7 million according to one study; and (4) provide more appropriate treatment by staff with expertise in management of SCD.¹⁶ The benefits of implementing day treatment clinics in Michigan are clear, however finding the necessary funding and resources to do so will be a challenge.
- ◆ **RESEARCH:** The overwhelming consensus in regards to SCD research is that much more could be done in our state to expand public health efforts and involvement. Strategic planning discussions determined that public health's role should be to establish a consortium for researchers, healthcare providers, public health professions, community groups, individuals and their families living with SCD and supporters, to work collaboratively in identifying barriers that are limiting creation, adoption and adherence to evidence-based screening recommendations, disease modifying therapies such as hydroxyurea, and best practices that help in the management of SCD. MDHHS can help to encourage research endeavors with qualitative and quantitative data and educational opportunities that will play a key role in empowering people and affecting change.

Based on these findings, the SCD Strategy Team developed goals and recommendations to raise awareness of this disease and to eliminate barriers to care.



The State Sickle Cell Disease Plan: Strategic Goals

- ◆ **Goal I: EDUCATION & AWARENESS** —To develop statewide multi-level messaging and communication strategies to increase awareness of SCD, disease-modifying medical therapies, and SCT.

A major and overwhelming theme through all components of the strategic planning process was the importance of increasing SCD and SCT awareness using a multi-tiered approach in order to maximize patient, family, provider, and community knowledge to improve health and quality of life. Although SCD was first recognized over 100 years ago, there is still a lack of general knowledge about the condition. Therefore, a major focus of the strategic plan will be to educate the general public, consumers, and health and human service professionals about the impacts of SCD and SCT.

STRATEGY: Increase activities with public and private organizations to raise awareness among targeted segments of the general and patient population about SCD/SCT and its implications.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD, CSHCS) SCDAA-MI, University of Michigan (UM)	(1.a) Develop a multi-level education strategy to inform the public at large about the burden of SCD; reach those who have, or are at risk for having SCT, and educate individuals with SCD to increase use of disease modifying therapies.	1
	(1.b) Work with SCDAA-MI and UM to develop targeted health education materials that include general SCD information, issues related to stigmatization, transition of care, pain management, genetic counseling, hydroxyurea, and the Family Medical Leave Act (FMLA).	Ongoing
MDHHS(LEGD, CSHCS), Children’s Hospital of Michigan (CHM), SCDAA-MI, Specialty Clinics, Medical Schools, Medicaid, Health Plans	(2.a) Work with SCDAA-MI, MCH programs, CSHCS, and the Family Center to connect services for individuals and families with special health care needs.	Ongoing
	(2.b) Distribute information to providers on lifecourse approaches to care transitions specifically related to SCD.	1,2
	(2.c) Include SCD information in Health Plan provider notices.	2,3
	(2.d) Explore the potential to collaborate with the Detroit Medical Center (DMC) and Children’s Hospital of Michigan (CHM) to establish a one year sickle cell fellowship program to exclusively train physicians, physician assistants, and nurses.	2,3
MDHHS (LEGD, CSHCS), SCDAA-MI	(3.a) Explore interest in developing a statewide SCD coalition; seek involvement from Michigan Health & Hospital Association, Michigan Primary Care Association, Legislative Black Caucus, school-based health centers, local health departments (LHD), health care providers, community-based organizations (CBOs), and consumers.	1
	(3.b) Promote public participation in rallies and walks to increase awareness.	Ongoing
	(3.c) Provide in-service training opportunities for MDHHS programs.	1,2,3

◆ **Goal I: EDUCATION & AWARENESS Continued**

STRATEGY: Increase capacity within MDHHS programs to incorporate SCD into public health programming.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD, CAHC, DOI, MIHP, WIC, CSHCS)	(4.a) Partner with MDHHS programs to deliver SCD and SCT education to their respective target populations (i.e., providers, pregnant women, young children and teenagers).	1
	(4.b) Post health education materials on the MDHHS website.	Ongoing
MDHHS (LEGD, MIHP)	(5.a) Explore opportunities to utilize the Home Visiting Network and Text for Baby to send medication (i.e., penicillin, hydroxyurea) and immunization reminder and/or notification alerts.	1
	(5.b) Review assessment and screening tools to incorporate applicable SCD questions to identify individuals with the condition and provide information and resources to reduce barriers.	1
	(5.c) Conduct outreach and provide informational materials to schools, colleges, and community-based programs (i.e., Federally Qualified Health Centers (FQHC), Child and Adolescent Health Centers) to increase awareness of SCD.	Ongoing
MDHHS (LEGD,DOI)	(6.a) Distribute and update special immunization recommendations to providers; add SCD-specific materials to the Adolescent Immunization Toolkit (AIM) and distribute to local health departments, Vaccine For Children Providers, and high risk clinics.	Ongoing
	(6.b) Develop immunization rate measures specific to SCD to monitor long-term trends.	1,2
	(6.c) Assess MCIR and HL7 messaging capacity to develop SCD notification module for providers.	1,2
MDHHS (LEGD, CAH)	(7.a) Provide in-service training on SCD via Grand Rounds once every 2 years for CAHC staff, and annually for school nurses/school staff.	2,3
	(7.b) Develop SCD education packet on family planning decisions for use in CAHC centers.	1
MDHHS (LEGD, FCCYSHCN)	(8.a) Develop a resource registry of SCD resources for MDHHS Family Center.	1
	(8.b) Increase awareness and utilization of the Family Center’s parent mentor matching program for SCD clients.	Ongoing
	(8.c) Increase awareness and utilization of the county-level Parent to Parent Support among families living with SCD.	Ongoing
	(8.d) Recruit parents of children with SCD to participate in mentor training to become mentors to other parents.	2,3

◆ **Goal II: TRANSITION —To develop and implement strategies for improving transition from pediatric to adult care.**

When young adults with SCD transfer their medical and non-medical care to adult services, they may feel uncomfortable and find it difficult to understand all that is expected of them, including the need to find new doctors, and navigate through insurance issues. A new emphasis on providing the necessary information and resources to help youth transition into the adult care setting will be pursued. Such initiatives will be of value not only to public health programs, but also to hematologists and primary care providers as they manage patient care in tandem with pediatric specialists to ensure successful transition.

STRATEGY: Increase resources and tools for specialty clinics, community-based organizations/clinics, and healthcare providers to utilize in order to systematically enhance the transition process for patients.

KEY PARTNERS	TACTICS	YEAR
MDHHS CSHCS, Specialty Clinics	(1.a) Identify the role of Children’s Multi-disciplinary Specialty Clinics (CMDs) in supporting clinic teams to address transition, including mechanisms for enhanced medical care using a team approach that includes a physician and a nurse/physician assistant at a minimum and if possible, a social worker and/or case manager.	2
	(1.b) Identify strategies and systems to aid specialty clinics in promoting transition from pediatric to adult care.	1
	(1.c) Identify approaches to reducing barriers surrounding establishment and utilization of transition care programs (i.e., creative financing solutions)	1,2
MDHHS CSHCS, SCDA-MI	(2.a) Identify and promote best practices for client, parent, and caregiver education to achieve competence in areas of independence and medical systems navigation.	1,2
	(2.b) Disseminate information to groups or forums where patients with SCD can share ideas and support one another.	Ongoing
SCDA-MI, Specialty Clinics	(3.a) Build patients’ knowledge and skills to understand and manage their health, healthcare needs, and to advocate for themselves.	Ongoing
	(3.b) Prepare parents to support patients’ independence.	Ongoing
	(3.c) Provide patient resources for ongoing health needs (i.e., reproductive health, genetic counseling, oral health, sickle cell complications that increase with age, maternal health needs).	Ongoing
MDHHS CSHCS, SCDA-MI,	(4.a) Explore interest in developing a working group designated for nurses, case managers, health educators and social workers to address transition tactics.	1
	(4.b) Identify strategies to facilitate communication among patients, families and providers.	Ongoing

◆ Goal II: TRANSITION Continued

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD, MIHP, CAH), SCDA-MI	(5.a) Develop a transition checklist for specialty clinics and primary care providers that includes pain management education and planning, peer mentoring, immunization education, trait education, genetic counseling, and resource awareness.	1
	(5.b) Create a targeted transition education initiative in CAHC program.	Ongoing
MDHHS (LEGD, CAH)	(6.a) Initiate a pilot project to identify primary care providers in Detroit area with the highest number of adolescent patients with SCD and partner to identify unmet needs.	2,3
	(6.b) Work with CAHC program providers to address the needs of students with SCD. - Conduct key informant interviews - Create transition education materials	2,3

◆ **Goal III: PROVIDER SHORTAGE** —To increase availability of providers who treat all aspects of SCD with an emphasis on increasing providers for adults.

Despite innovations in medical care and improved survival among individuals with SCD, clinical management for the condition continues to pose a substantial challenge for patients, families, and providers. SCD requires a significant commitment of medical resources including pediatric and adult hematology, nursing and other sub-specialists. National studies show an increase in the morbidity rate after transitioning to adult care due at least in part to a lack of comprehensive programs for adults with SCD.⁸ In Michigan, there is a clear lack of adult providers for either primary or subspecialty care. With only one adult clinic in Detroit devoted to treating adult patients, it is unclear where remaining adult patients receive care. These tactics aim to address the burden of provider shortages on the patient and health care system.

STRATEGY: Increase the number of providers caring for sickle cell patients in Michigan.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD)	(1.a) Investigate medical school scholarship opportunities and loan repayment policies for physicians to practice in underserved areas (i.e., primary care physicians for children, hematologists for adults).	2
MDHHS (LEGD), SCDAA-MI	(2.a) Explore existing educational tools and identify training gaps; explore sources to increase specialty education and training opportunities for providers. (2.b) In collaboration with the Midwest region HRSA grant initiative, organize education and training opportunities for health care providers treating patients with SCD. (2.c) Share NHLBI Evidence-Based Management of SCD Guidelines with the primary care provider of every baby diagnosed with SCD. (2.d) Collaborate with HemQIC and Michigan Quality Improvement Consortium to identify activities, training opportunities, and incentives that will be needed to increase the number of providers treating patients with SCD. (2.e) Identify and apply for applicable funding opportunities.	2 Ongoing 2,3 Ongoing Ongoing
MDHHS (LEGD, CSHCS)	(3.a) Quantify the ratio of pediatric and adult hematologists in relation to target population residential location to demonstrate the need to increase provider capacity. (3.b) Assess enrolled CSHCS subspecialty providers caring for children and youth with SCD to determine gaps in provider mix compared to other disorders. (3.c) Identify opportunities to foster specialist and primary care provider joint case management through use of telemedicine.	1,2 2 2,3
MDHHS (LEGD, Health Plans, Medicaid), HemQIC	(4.a) Review Medicaid Health Plan Guidelines and compare to NHLBI Evidence-Based Management of SCD Guidelines. Develop Michigan-specific guidelines that reflect national standards.	1,2
MDHHS (LEGD), HemQIC, Michigan Nurses Association	(5.a) Develop a SCD pain training module for nurses and provide continuing education credits (CEUs).	2

◆ **Goal IV: PSYCHOSOCIAL / MENTAL HEALTH SUPPORT** —To increase recognition of the need to address psychological issues and provide mental health support for individuals living with SCD and their families.

Psychological complications in patients with SCD mainly result from the impact of pain and symptoms on their daily lives and society’s attitudes towards them.¹⁷ Studies have shown that the most frequent psychological problems encountered include increased anxiety, depression, social withdrawal, aggression, poor relationships and poor school performance.¹⁷ A few case reports also indicated high levels of parental anxiety, overprotection, excessive feelings of responsibility and guilt. These issues are further complicated by the social, economic, and healthcare disparities experienced by many patients with SCD. Adequate psychological support could help to improve an individual’s quality of life. In addition to clinical treatment of the disease, assessment of psychological factors is important.

STRATEGY: Increase partnerships and resources for mental/behavioral health providers to decrease rates of psychiatric morbidity in the SCD population.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD, CSHCS, MCH, Office of Recovery Oriented Systems of Care)	(1.a) Identify relevant programs and points of contact within MDHHS to address psychological and mental health needs.	2
	(1.b) Identify existing psychological needs assessment tools suitable for patients and families affected by SCD.	2
	(1.c) Identify relevant programs to support reproductive counseling and prenatal education.	1
	(1.d) Promote access to substance abuse treatment services for individuals with SCD as needed.	Ongoing
MDHHS (LEGD, CSHCS), SCDA-MI, Specialty Clinics	(2.a) Promote alternative and/or holistic therapies for acute pain management.	Ongoing
MDHHS (LEGD, WIC, Community Services Bureau), SCDA-MI	(3.a) Identify relevant programs and points of contact to promote awareness and understanding of SCD among mental health providers, therapists, and educators.	2,3
	(3.b) Partner with the MDHHS Community Services Bureau to Improve awareness and identify community resources that will provide assistance to individuals living with SCD.	Ongoing
MDHHS (LEGD), SCDA-MI, HemQIC, Specialty Clinics	(4.a) Share guidelines for treating emergency room patients who are both in pain and drug-seeking.	1,2
	(4.b) Identify partnerships to develop training for emergency room staff to treat patients who are in pain and potentially drug-seeking.	2,3

◆ **Goal V: MEDICATION ADHERENCE — To develop protocols for improving medication adherence in accordance with the NHLBI Evidenced-Based Management of SCD Guidelines.**

In Michigan, it is mandatory to screen every newborn for SCD in order to facilitate the start of prophylactic penicillin and anticipatory guidance prior to four months of age. In addition to daily administration of penicillin until at least five years of age, children and adults with SCD often require other daily medications such as folic acid, hydroxyurea, and oral iron chelation therapy to decrease the risk of complications. Given the high expectation on this patient population to take daily medication to prevent complications of the disease, understanding and improving the rate of adherence is an important component of patient care.

STRATEGY: Increase understanding of patient motivating factors for medication adherence.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD, CSHCS, Medicaid, Managed Care), SCDAAs-MI, HemQIC	(1.a) Characterize barriers to patient adherence for recommended treatments.	1,2
	(1.b) Collaborate with partners to identify and promote initiatives to overcome barriers and improve self-managed adherence.	Ongoing
	(1.c) Support education to promote adoption of model treatment practices and disease modifying therapies.	Ongoing
	(1.d) Assess the effectiveness of task-focused communication technology (i.e., Glocap, VOICE Crisis Alert) in improving adherence.	2,3
MDHHS (LEGD), SCDAAs-MI, Specialty Clinics	(2.a) Promote uniform practice guidelines among specialty providers; Use an established prescribing and monitoring protocol.	2,3
MDHHS (LEGD), SCDAAs-MI, HemQIC, Specialty Clinics	(3.a) Conduct literature review to identify myths and strategies to dispel myths and promote facts about SCD patient attitudes, behaviors, and cultural competence.	Ongoing
	(3.b) Identify and disseminate written patient materials with plain language information.	Ongoing

◆ **Goal VI: DAY TREATMENT CLINICS — To improve acute care in the emergency room or alternative settings.**

Recurrent episodes of pain are the most consistent expression of SCD. Pain management involves care of both acute and chronic pain. One of the important interventions that has improved the care of patients with SCD is the institution of a day treatment clinic where acute episodes of pain can be managed without resorting to emergency room care. Day treatment clinics offer many advantages: 1) the majority of patients return home the same day; 2) treatment can begin within minutes of arrival; 3) staff are familiar with the patients and their analgesic requirements for pain control; 4) reduced need for hospitalization and fewer visits to the emergency department; and 5) reduced cost of care.

STRATEGY: Partner with hospital systems to promote opportunities, training, and best practices to increase patient satisfaction in the emergency room or alternative settings.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD, CSHCS), SCDA-MI, HemQIC	(1.a) Identify best practices for acute pain management in various health care settings.	Ongoing
	(1.b) Identify strategies for the successful dissemination and implementation of best practices in collaboration with partners.	Ongoing
	(1.c) Compile and distribute best practice recommendations and algorithms that can be adapted for use in various acute settings.	2,3
MDHHS (CSHCS), Specialty Clinics	(2.a) Promote continuous quality improvement projects to reduce ED wait time to initial administration of pain medication.	2,3
MDHHS (LEGD, CSHCS, Medicaid, Health Plans), Specialty Clinics, SCDA-MI	(3.a) Assess the feasibility and cost-benefits of establishing additional day treatment clinics.	2,3
	(3.b) Investigate public and private funding sources to support day treatment programs (i.e., DMC, Hurley Hospital).	2,3

◆ **Goal VII: RESEARCH — To establish a Michigan SCD Consortium to coordinate public health research efforts within the state of Michigan.**

Over the past several decades, scientists and doctors have learned a great deal about SCD. They know its cause, how it affects the body, and how to treat many of its complications. Thanks to improved treatment and care, people who have SCD are now living into their 40s, 50s or longer. A Michigan SCD Consortium would bring together leading hematologists, public health professionals, people with SCD and their families, to find solutions to the many problems posed by SCD. Increased investment in the development of research questions will be key to achieving this goal.

STRATEGY: Increase public health capacity to facilitate research opportunities in Michigan.

KEY PARTNERS	TACTICS	YEAR
MDHHS (LEGD)	(1.a) Develop a consortium to coordinate public health research efforts; establish panel experts in SCD, appropriate specialists—emergency medicine, pain medicine, psychology, primary care providers, patients, public health, hematology.	2,3
	(1.b) Facilitate periodic stakeholder meetings and symposia to share progress on the MDHHS SCD strategic plan, management updates, research priorities and quality assurance outcome measures for discussion and consensus development.	2,3
	(1.c) Review core hemoglobinopathy indicators annually.	Ongoing
	(1.d) Update and distribute SCD data on a regular basis.	Ongoing
MDHHS (LEGD), Consortium	(2.a) Identify and promote implementation of a SCD research agenda for public health	2,3
	(2.b) Review available qualitative and quantitative data relating to SCD management.	2,3
	(2.c) Promote public health research initiatives to improve health outcomes for individuals with SCD.	Ongoing



Next Steps: Approach to Implementation and Opportunities For Collaboration

This plan reflects the knowledge, skills, expertise and passion of a dedicated group of action-oriented agencies and individuals. The MDHHS LEGD will continue to lead implementation of the plan with participation and collaboration from internal public health programs, the SCDA-MI, HemQIC, Specialty Clinics, community groups, and individuals living with SCD. Meetings with the MDHHS Strategy Team will be conducted at least twice per year to monitor plan progress. In Year 1 the Strategy Team will develop an evaluation plan and then monitor progress of program improvement and decision making.

Many aspects of this plan are already being implemented. As Michigan's SCD initiatives move forward, the department is well poised to deliver services and support to those with SCD and/or their families through existing programs. The LEGD will also continue participating in opportunities and initiatives that complement and coordinate with our goals including:

- ◆ Participation in the HRSA 3-year Sickle Treatment and Outcomes Research in the Midwest (STORM) funding opportunity led by the Cincinnati Comprehensive Sickle Cell Center/Children's Hospital Medical Center, along with state-level partners in Indiana, Illinois, Minnesota, Ohio and Wisconsin. The overarching goal of this project is to improve the quality of life and health outcomes for all Midwesterners with SCD and to bring together hematology and primary care communities to collaborate with patients and families. STORM priorities are to: 1) increase the number of providers caring for sickle cell patients in the region; 2) increase the number of providers prescribing hydroxyurea in the region; and 3) increase the number of sickle cell patients receiving treatment from a knowledgeable provider in the region.
- ◆ Continued involvement with the Region 4 Midwest Genetics Collaborative Hemoglobinopathies Workgroup to provide a forum for families, public health, and clinical providers to share best practices and models for improving NBS, follow-up and genetic care coordination for children with hemoglobinopathies and their families. Workgroup activities have included completing a regional baseline assessment of current practices for SCT follow-up; developing best practices for the notification and education for families of children identified with SCD through NBS; developing SCT family education materials; and adapting the medical home guidebook "Partnering with your Doctor" for families of children with SCD.
- ◆ Building upon surveillance initiatives to monitor long-term patient follow-up indicators (i.e. antibiotic prophylaxis) over time; measuring long-term trends in quality of care and outcomes specific to the SCD population of Michigan; and creating benchmarks for comparison with national hemoglobinopathy standards.
- ◆ Collaboration with the SCDA-MI and HemQIC to utilize a 56-question health status assessment (HSA) survey developed to measure access to care, disease complications, health care use, financial status, and education status among people in Michigan with SCD. Over the last several years the HSA has provided self-reported information on patients, in order to learn more about Michigan's population with SCD, provide feedback on education initiatives provided by the SCDA-MI, and facilitate conversation among patients and their families about important health issues related to SCD. The HSA will continue to be included as part of regular follow-up for all SCD patients detected via NBS.

References

1. Petrea-Cober M, Phelps S. "Penicillin Prophylaxis in Children with Sickle Cell Disease." *The Journal of Pediatric Pharmacology and Therapeutics*. 2010 Jul-Sep; 15(3): 152-159.
2. Penn State Hershey Milton S. Hershey Medical Center. "Sickle Cell Disease Highlights." http://printer-friendly.adam.com/content.aspx?productId=10&pid=10&gid=000058&c_custid=758. Accessed April 15, 2015.
3. Fitzhugh CD, Lauder N, Jonassaint JC, Telen MJ, Zhao X, Wright EC, Gilliam FR, De Castro LM. Cardiopulmonary Complications Leading to Premature Deaths in Adults with Sickle Cell Disease. *American Journal of Hematology*. 2010 Jan;85(1): 36-40.
4. Centers for Disease Control & Prevention. What You Should Know About Sickle Cell Trait. http://www.cdc.gov/ncbddd/sicklecell/documents/SCD%20factsheet_Sickle%20Cell%20Trait.pdf. Accessed January 2015.
5. Centers for Disease Control & Prevention. How Sickle Cell Trait Is Inherited. <http://www.cdc.gov/ncbddd/sicklecell/traits.html>. Accessed May 23, 2014.
6. Stottlemeyer R, Kleyn M, Garcia E, Smith D, Bach J, Young W, Lyon-Callo S. Sickle Cell Disease: 27 Anniversary Newborn Screening Surveillance Report. Michigan Department of Community Health. October 2014. http://www.michigan.gov/documents/mdch/RuSH_Final_Draft_11.07_473558_7.pdf. Accessed February 1, 2015
7. Michigan Department of Health & Human Services. Michigan Newborn Screening Program Annual Report: http://michigan.gov/documents/mdch/Annual_Report2013_final_474204_7.pdf. Accessed June 12, 2015.
8. DeBaun, MR, Telfair J. "Transition and Sickle Cell Disease." *Journal of American Academy of Pediatrics*. Originally published online October 1, 2012. DOI: 10.1442/peds.2011-3049. <http://pediatrics.aappublications.org/content/early/2012/09/26/peds.2011-3049.full.pdf>. Accessed June 17, 2015.
9. Jenerette, CM, Brewer C. "Health-Related Stigma in Young Adults with Sickle Cell Disease." *Journal of National Medical Association*. 2010 Nov; 102(11): 1050–1055.
10. National Heart Lung and Blood Institute, "Management of Sickle Cell Disease," *NIH Publication Number 02-2117*, Reprinted June 2002, p. 55.
11. Epping, A. S., Myrvik, M. P., Newby, R. F., Panepinto, J. A., Brandow, A. M., & Scott, J. P. (2013). Academic attainment findings in children with sickle cell disease. *Journal of School Health*. 2013 August; 83(8), 548-553.
12. Patient-Centered Outcomes Research Institute. Patient-Centered Comprehensive Medication Adherence Management System to Improve Effectiveness of Disease Modifying Therapy with Hydroxyurea in Patients with Sickle Cell Disease. <http://www.pcori.org/research-results/2013/patient-centered-comprehensive-medication-adherence-management-system-improve>. Accessed June 17, 2015.
13. Kofi AA, Feyijimi EE, Olu A. "Psychosocial Impact of Sickle Cell Disorder: Perspectives from Nigeria Setting." *Globalization and Health*. <http://www.globalizationandhealth.com/content/6/1/2>. Accessed June 17, 2015.
14. Bhatia M, Walters MC: "Hematopoietic Cell Transplantation for Thalassemia and Sickle Cell Disease: Past, Present and Future." *Bone Marrow Transplant* 2008, 41:109-17.
15. De Montalembert M: "Management of Sickle Cell Disease." *BMJ* 2008, 337:a1397.
16. Benjamin LJ, Swinson GI, Nagel RL. Sickle Cell Anemia Day Hospital: An Approach for the Management of Uncomplicated Painful Crises. *Blood*. 2000 Feb 15; 95(4): 1130-6.
17. Kofi A Anie. "Psychological Complication of Sickle Cell Disease." *British Journal of Haematology*. Volume 129, Issue <http://onlinelibrary.wiley.com/doi/10.1111/j.1365-2141.2005.05500.x/pdf>. Accessed June 15, 2015.
18. National Heart, Lung, and Blood Institute. A Century of Progress: Milestones in Sickle Cell Disease Research and Care. <http://www.nhlbi.nih.gov/files/docs/public/blood/Tagged2NHLBISickleCellTimeline.pdf>. Accessed April 1, 2015.
19. Centers for Disease Control & Prevention. Sickle Cell Disease Data & Statistics. <http://www.cdc.gov/ncbddd/sicklecell/data.html>. April 1, 2015.
20. American Society of Hematology. Sickle Cell Trait. <http://www.hematology.org/Patients/Anemia/Sickle-Cell-Trait.aspx>.



Appendix: Key Milestones of Sickle Cell Disease

The past 100 years of sickle cell research have resulted in landmark discoveries that ushered in the era of molecular genetics.¹⁶ In the United States, SCD affects an estimated 90,000 to 100,000 people, the majority of whom are African Americans.¹⁷ All states screen newborns for SCD. The condition occurs in approximately one out of every 500 African American births and one out of every 36,000 Hispanic American births.¹⁸ In addition, more than 2 million people in the United States have SCT.¹

1910 ~	Dr. James B. Herrick publishes first description of sickled cells in blood sample from 20 year old dental student from Grenada. Term “sickle cell anemia” coined based on paper.
1933 ~	Scientists test 2500 African Americans in Memphis; determine SCT and SCD are separate entities.
1940 ~	Researchers suggest exchange of oxygen for carbon dioxide occurring in small blood vessels may cause red RBCs to sickle and block blood vessels.
1948 ~	Dr. James Neel suggests low concentration of sickled cells in blood from newborns with SCD due to high level of fetal Hgb in their RBCs.
1949 ~	Dr. Linus Pauling reveals that SCD is due to abnormal Hgb protein molecule. Term “molecular disease” coined.
1949 ~ 1950	Inheritance of SCD independently described by two teams. Sickle cell genes needed from both parents to produce SCD. Receiving gene from one parent produces SCT.
1954 ~	SCT found to protect against malaria. Finding explains why the prevalence of the sickle gene in Africa corresponds with regions where malaria is a major cause of death.
1957 ~	Scientists show abnormality of sickle Hgb due to amino acid substitution in protein, making SCD the first genetic disorder whose molecular basis is known.
1972 ~	National Sickle Cell Anemia Control Act provides for establishment of voluntary SCD screening; counseling; public and professional education; and research and training in diagnosing, treating, and controlling disease; A milder variation of SCD was found in Saudi Arabia associated with increased levels of fetal hemoglobin. Findings suggested increasing fetal Hgb levels could offer treatment target.
1972 ~	Sickle Cell Detection Center established in Detroit, Michigan.



Appendix: Key Milestones of Sickle Cell Disease cont.

1974 ~	Feasibility of NBS for SCD demonstrated. Method developed for prenatal diagnosis by sampling fetal blood from the umbilical vein.
1978 ~	New York becomes first state to screen for SCD; Prenatal method to diagnose SCD using DNA samples reported. The NHLBI launches multicenter study with 4,000-plus individuals from newborns to age 70. First study to document clinical course of disease from birth to adulthood.
1984 ~	Several teams independently demonstrate that hydroxyurea increases fetal Hgb levels. Bone marrow transplant performed to treat a child with leukemia also cures the child's SCD.
1986 ~	An NHLBI study shows penicillin as a preventive measure in children with SCD 3 months to 5 years old can reduce the incidence of Streptococcus pneumonia infection, a major cause of childhood death, by 84 percent. Practice later becomes widely adopted.
1987 ~	National Institutes of Health Consensus Development Panel Recommends screening all U.S. newborns for SCD and giving penicillin to all affected infants by 3 months of age.
1987 ~	Michigan legislature adds SCD to the state NBS panel and state health department establishes contract with the SCDA-MI to coordinate follow-up services for babies detected with SCD.
2014 ~	An expert panel of the NHLBI released new guidelines for managing SCD.
2014 ~	MDHHS-LEGD released the Sickle Cell Disease Newborn Screening Surveillance Report celebrating 27 years of sickle cell screening.



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2015—2018

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