Report to the General Assembly

Public Act 102-0004 (410 ILCS 460/131-25)

Sickle Cell Statewide Impact and Surveillance Program

State of Illinois
JB Pritzker, Governor

Illinois Department of Public Health
Office of Health Promotion
Health Assessment and Screening Section
535 West Jefferson Street
Springfield, Illinois 62761-0001
July 9, 2024

In partnership with
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To the Members of the General Assembly:

I am pleased to share this report updating our progress on the Sickle Cell Statewide Impact and Surveillance Program. This report is the culmination of months of work by professionals within the Illinois Departments of Public Health and Healthcare and Family Services, along with colleagues and advocates around the state working on behalf of those dealing with sickle cell disease (SCD).

SCD is an inherited disorder of red blood cells, disproportionately impacting Black and Latino populations. It can be a painful condition, one that is linked to serious and potentially debilitating medical complications.

The information contained in this report serves as a sad reminder of the tremendous toll that SCD takes on patients and families. It means frequent trips to emergency departments and hospital admissions. It means recurring difficulties in finding qualified caregivers, negotiating insurance coverage challenges, and even finding transportation to obtain necessary services. And all too often, it means a premature death.

The data is sobering, but it is also essential to our mission to identify resources and solutions that can help SCD patients and their families get the help they need to live longer, happier, and healthier lives.

Through the foresight of the General Assembly and Governor JB Pritzker, the Sickle Cell Prevention Care and Treatment Act entrusts us with identifying individuals impacted by SCD and connecting them to critical resources. This report takes us a significant step closer to meeting those goals. The information contained here will help us, in collaboration with the health care community and with the support of our legislative partners, to break down the geographic, financial, and bureaucratic barriers to care and to help people across the state get the medical, psychological, and social assistance they need to live and thrive after an SCD diagnosis.

I thank you for your efforts to address this issue and support the work of professionals around the state to help persons and families impacted by SCD. Together, we can and will chart a path for brighter days and better health ahead.

Yours in good health,

Sameer Vohra

Sameer Vohra, MD, JD, MA
Director

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The Illinois Department of Public Health dedicates this report to all Illinoisans who are coping with sickle cell disease with courage and strength, and to their loved ones and families.

We hope our efforts to better understand the challenges facing these individuals as they navigate the health care system and their daily lives will prevent suffering by supporting systemic changes that promote health and health equity for all.

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Acknowledgment
Elizabeth Grace Borto, Intern
Created the images used in the "Fundamentals of Sickle Cell Disease" section.

Purpose of the Report

Public Act 102-0004 created the *Sickle Cell Prevention, Care, and Treatment Program Act and Sickle Cell Chronic Disease Fund*. The act requires that the Illinois Department of Public Health (IDPH) conduct a study to determine the prevalence, impact, and needs of individuals with sickle cell disease (SCD) and sickle cell trait in Illinois (410 ILCS 460/131-25).

Specific requirements for the report are laid out in the Illinois Administrative Code, Title 77 Section 663.200, which include:

1. Develop and establish the Illinois Sickle Cell Impact Surveillance System (ISCISS) to assess and monitor the impact of sickling disorders on Illinois residents.
2. Determine:
   a. The prevalence, by geographic location, of people diagnosed with sickle cell disease and sickle cell trait in Illinois.
   b. The availability and affordability of screening services for sickle cell trait in Illinois.
   c. The location and capacity for the treatment of sickle cell disease and sickle cell trait.
   d. The unmet medical, psychological, and social needs of people in Illinois with sickle cell disease.
   e. The underserved areas of Illinois for the treatment of sickle cell disease.
3. Recommendations for actions to address any shortcomings in the state identified under this section.
4. Make the same determinations for additional hemoglobinopathies which may impact the health and well-being of affected persons.

Acknowledgment of Grant Funding

The Sickle Cell Prevention Care and Treatment Act [410 ILCS 460] established a grant program to fund services for adults with sickle cell disease, adolescents in transition, and for educational programs concerning the disease. Funding may support:

- Assisting in the development and expansion of care for the treatment of individuals with sickle cell disease, particularly for adults and adolescents transitioning to adult services.
- Developing new or enhancing educational and outreach programs to increase access to health care and support services for individuals with sickle cell disease and sickle cell trait and their families.
- Establishing or enhancing sickle cell disease infusion centers.
- Increasing access to mental health resources and pain management therapies for individuals with sickle cell disease.
- Providing counseling to any individual, at no cost, concerning sickle cell disease and sickle cell trait, and the characteristics, symptoms, and treatment of the disease.

The IDPH Sickle Cell Prevention Care and Treatment Grant Program accepted its first competitive applications for fiscal year 2025. Ongoing work will depend on the Illinois General Assembly continuing to set aside funds for the Sickle Cell Chronic Disease Fund.

Executive Summary

Introduction

Sickle cell disease (SCD) is a life-long inherited disorder affecting red blood cells that results in significant complications, negatively affecting the life of the individual, and requiring frequent specialized care. This condition primarily affects Black or African American and Hispanic or Latino populations, and it has long been recognized that many people with SCD struggle to receive the care that they need. In response, the Illinois legislature, through Public Act 102-0004, has laid out measures to better understand the current challenges and barriers to care through the preparation of this biennial report and the provision of funds to support improvements in care.

For this inaugural report, IDPH partnered with the Illinois Department of Healthcare and Family Services (HFS) to review public health and Medicaid data for people with SCD. In addition, IDPH conducted a survey of health care providers with expertise in SCD, who routinely interact with the IDPH Newborn Screening Program.

Initial Findings

- The initial estimate for the number of people living with SCD in Illinois is at least 6,000, based on Medicaid claims data.
- Most people with SCD in Illinois live in urban areas, primarily in the Chicago metropolitan area where the majority of their health care is provided. SCD health care specialists in St. Louis are another focus for care.
- Illinois data reveal the high negative impact of this disease, mirroring national trends.
- People with SCD in Illinois often require visits to the emergency department and are frequently admitted to the hospital for severe complications of the disease beginning in early adulthood.
- People with SCD are dying at a much younger age than the Illinois population, with death occurring in middle age. Few people with SCD are reaching the age of 65 years.
- Responding to a survey, SCD health care providers listed significant barriers to patients accessing needed care. The top three identified barriers were 1) transportation challenges impeding patients' access to necessary health care services, 2) lack of available specialists and difficulty accessing specialized care, and 3) complications with insurance coverage, including referrals and provider networks.

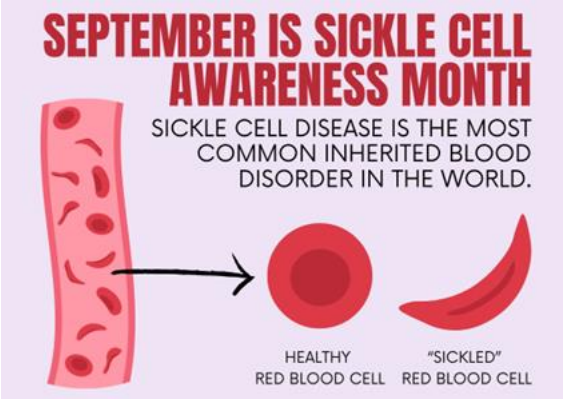
Plans for future reports

This first report summarizes the initial steps that IDPH has taken to understand the current system for providing health care to people with SCD. The data clearly show the significant negative impact of SCD on the lives of affected Illinoisans. Further investigation is urgently needed to better understand the many factors that are contributing to people with SCD not receiving evidence-based, clinically recommended care.

IDPH is dedicated to improving outcomes for people with SCD. In the coming months, IDPH will engage health care providers, people who have SCD, and advocacy groups, such as the Sickle Cell Disease Association of Illinois (SCDAI), to further understand the barriers that exist and develop recommendations for action.

IDPH will address these issues by collaborating with entities that provide care and services to affected individuals. The IDPH Sickle Cell Prevention Care and Treatment Grant Program will be integral to this work.

Fundamentals of Sickle Cell Disease

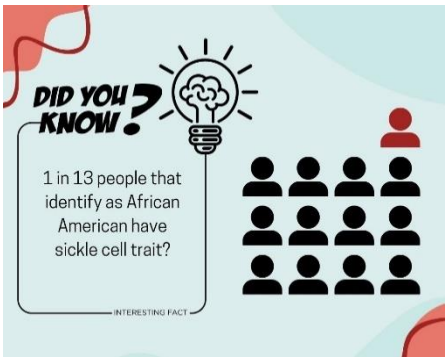


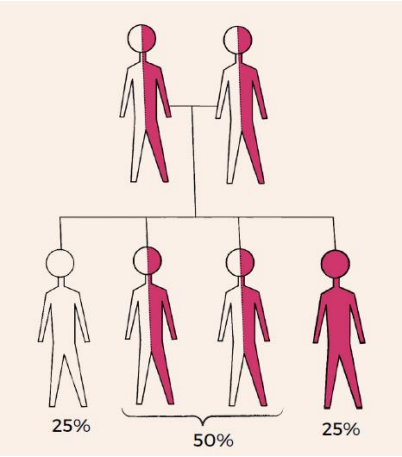
Sickle cell disease (SCD) is the most common inherited blood disorder in the United States. People with SCD have a genetic mutation in hemoglobin, the protein that carries oxygen through the body. Red blood cells normally are flexible enough to move easily through blood vessels. In SCD, however, the abnormal hemoglobin causes red blood cells to become sickle-shaped, hard, and sticky, leading to blockages of blood vessels.

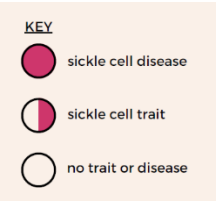
The blocked blood flow can affect every organ in the body and cause serious problems, including episodes of severe pain, stroke, heart disease, kidney disease, lung disease, and loss of vision. People with sickle cell disease can start having episodes of severe pain and other complications after the first few months of life.

Stressful conditions, including infection, dehydration (not enough fluids in the body), cold weather, emotional stress, pregnancy, or alcohol intoxication, can trigger symptoms of sickle cell disease.

In the United States, sickle cell disease is most common in African Americans. About 7% of African Americans (about 1 in 13) have sickle cell trait and 1 in 365 have SCD.[1] It also occurs in people from the Caribbean, Mediterranean, South and Central America, and the Middle East.







Inheritance

SCD is passed down through families. Most people have two normal hemoglobin genes, but a person with sickle cell disease has two abnormal hemoglobin genes, one from each parent. People with sickle cell trait have one normal gene for hemoglobin and one abnormal gene.

The parents of a child with a sickle cell disease usually have sickle cell trait and are unaffected, healthy carriers. With each pregnancy, carrier parents have a 25% chance of having a child with two abnormal hemoglobin genes, resulting in SCD. Carrier parents have a 50%chance of having a child who is an unaffected carrier and a 25% chance of having an unaffected, non-carrier child. These risks hold true for each pregnancy.

It is also possible for people with one sickle cell gene, to inherit a different abnormal hemoglobin gene (such as Hb F, Hb C, Hb E, thalassemia trait) from

[1] Centers for Disease Control and Prevention Data and Statistics on Sickle Cell Disease

their other parent. These individuals may develop sickle cell disease conditions that vary, causing minor to serious health problems.

Treatment

SCD is a complex, life-long illness that can cause devastating complications and requires a multi-disciplinary team approach to providing care. Because of advances in treatment, most children with SCD are now living into adulthood. Treatment of SCD in early childhood includes administration of prophylactic (dosing of medication to prevent infection) penicillin up to 5 years of age to prevent bacterial infections, especially those that cause pneumonia and meningitis (brain infections); hydroxyurea to reduce sickling and prevent complications; and pain relief medication. Blood transfusions are often also needed. While bone marrow transplantation and gene therapy may cure sickle cell disease, these are not easily accessible at present.

Illinois Department of Healthcare and Family Services (HFS) Medical Benefits (*Illinois Medicaid*)

HFS is a key partner for IDPH in the assessment of the health care needs of people living with SCD because the department administers *Illinois Medicaid*, a federal-state program that provides affordable health insurance coverage for many Illinoisans who have the disease.

According to the Centers for Medicare & Medicaid Services (CMS), 50 – 60% of all individuals with SCD are covered by Medicaid, "which puts Medicaid in a critical role for mitigating poor health outcomes and cost burden for patients."[2] Illinois has a higher than national prevalence of SCD in the Medicaid beneficiary population.[3]

In September 2023, CMS released a Sickle Cell Disease Action Plan outlining strategies for improving access to care, quality of care, engagement, data analysis, and addressing equity.

IDPH Sickle Cell Newborn Screening Program

Newborn screening for sickle cell disease is crucial because referral to specialized care and treatment starting in the first few months of life can significantly reduce the severe childhood consequences of the disease.

Illinois began screening newborns for sickle cell disease in 1989. To perform newborn screening, a small quantity of blood is collected from the baby's heel 24-48 hours after birth and placed on special cards (also called blood cards) that are shipped by overnight courier to the IDPH Newborn Screening Laboratory.

Newborns are screened for more than 40 conditions, and the results are reported to the newborn's primary care provider. If a newborn has an out-of-range (abnormal) SCD result, it requires referral for specialized care involving clinical evaluation, confirmation of diagnosis, plans for medical management, and genetic and risk

[2] Sick-Cells_Medicaid-Access-and-Landscape-Review_Final-Report 2022/08.pdf
[3] CMS Infographic Beneficiaries with Sickle Cell Disease, September 2020

counseling. The IDPH Newborn Screening Program staff ensure that the primary care provider has made that referral and verify with the specialist the results of the initial evaluation.

An important IDPH Newborn Screening Program activity related to sickle cell disease screening is the Hemoglobinopathy Collaborative, a bi-monthly conversation between health care providers, SCD advocates/educators, and IDPH to facilitate communication and information sharing to improve the outcomes and care for patients with sickle cell disease.

In addition, the IDPH Newborn Screening and Genetics Sickle Cell Follow-Up Grant Program helps fund pediatric hematology centers to ensure the availability of statewide services for families who have a newborn or child with sickle cell disease.

Newborn screening methods, processes, and systems are continuously evolving. The Data Source and Methods Section presents selected IDPH Newborn Screening Program data. Changes in the numbers of individuals identified through newborn screening over the years are due to natural change, population shifts, and changing birth rates.

Genetic counseling services are recommended for individuals with sickle cell disease and for those who carry abnormal hemoglobin, particularly concerning future pregnancies. These individuals may have questions about the disorders that are best answered by hematology specialists and genetic counselors.

More information about the IDPH Newborn Screening Program can be found at:
https://dph.illinois.gov/topics-services/life-stages-populations/newborn-screening.html

Data Sources and Methods

Data is aggregated for January 1, 2016 – December 31, 2022, unless otherwise noted.

IDPH Newborn Screening Program

IDPH maintains a database of all NBS specimens submitted to the state laboratory for analysis and results. Information regarding individuals, specimens, and their respective screening outcomes was queried from the NBS program data management system for all babies screened from 2016-2022 who had a positive (abnormal) sickle cell screening result. Any newborn in this group with more than one newborn screening specimen or positive sickle cell screening result was only included once in the NBS data included in this report.

HFS Medicaid Billing

Medicaid claims were extracted from a list of individuals who had a primary diagnosis of sickle cell disease (i.e., code beginning with D57, excluding D573) from January 1, 2016, through December 31, 2022. Appendix C includes a list of the International Classification of Diseases, Tenth Revision (ICD-10) codes used to find cases of sickle cell disease.

Illinois Vital Records System Mortality Data

The Illinois Vital Records System (IVRS) contains birth and death records and is managed by the IDPH Division of Vital Records.

Funeral directors statewide are required by law to submit electronic death certificates to the IDPH Division of Vital Records, while medical certifiers (coroners/medical examiners, attending physicians, physicians in charge) determine the cause of death. Mortality data were queried for 2016 – 2022, and sickle cell disease-related deaths were defined as those in which sickle cell disease was reported as the underlying or contributing cause of death.

Hospital Admission Data

This report analyzed a subset of IDPH Hospital Discharge Data, including individuals with a diagnosis of sickle cell disease (coded as D57, excluding D57.3) between January 1, 2016, and December 31, 2022. The Division of Patient Safety and Quality collects patient-level discharge data from all Illinois hospitals and ambulatory surgical treatment centers. Programs within IDPH use discharge data for surveillance, health care planning, and public reporting.

Survey of Health Care Providers Who Interact with the Newborn Screening Program

Participants were recruited using a multi-step process. Participation was voluntary and confidential.

1. **Initial List Compilation:** The 19 SCD providers who routinely interact with the IDPH newborn screening program were invited to participate in the survey.
2. **Verification via Phone Calls:** Phone calls were made to reconfirm the contact details and SCD care involvement of providers on the compiled list.
3. **Data Collection:** Each provider received a customized email explaining the survey's objectives and its importance to enhancing SCD care in Illinois, a link to the Qualtrics survey, and the contact information of a designated person at IDPH for any questions or additional information. Participants were given two-weeks to complete the survey. Two follow-up reminder emails were sent. The survey was conducted between April 26 and May 6, 2024.

Survey of SCD Providers Who Work with the Newborn Screening Program Conducted April 26, 2024 – May 6, 2024 (Tables 1 and 2)

SCD Provider Survey Participant Overview Represented diverse specialties and areas of the state		
13 of 19 invited providers participated. Geographically located across Illinois.	Included physicians, nurses, genetic counselors, administrators, and social workers.	Worked in hospitals, clinics, a community-based organization, and universities.
Experience ranged from less than five years to more than 30 years in SCD care.	Provided care for pediatric and adult patients. Most cared for patients in the 0-10 and 11-20 age groups	The number of SCD patients varied widely, with some providers managing more than 50 patients.

Highly Effective Strategies to Overcome Barriers to Care: Quotes from Providers	
Identified Strategies	**Description**
Outreach Clinics	"One of the foremost strategies is the establishment of outreach clinics, which have brought medical services closer to patients."
Quality Improvement (QI) **Community Health Workers** **Shared Decision-Making**	"Emphasis on Quality Improvement (QI) work and the involvement of community health workers as integral team members have been crucial. These efforts include shared decision-making and involving patients in best practices, particularly in forming new adult clinics."
Social Determinants of Health	"The establishment of these clinics as trusted resources has addressed many social determinants of health."
Collaborate with Community Agencies **Individualized Care Plans** **Standards of Care**	"Collaborations with community agencies to develop individualized care plans and create standards of care that significantly improve patients' lives. The team also conducts regular conversations with parents, featuring speakers to identify the type of assistance needed and how best to provide it. This collaborative approach has proven effective in tailoring care to meet individual needs."
More Effective Treatment	"The use of treatment-modifying agents has also been effective in reducing the complications associated with SCD."
Educational Liaison **Individualized Education Programs (IEPs)**	"Educational liaison to help patients with school attendance and access to Individualized Education Programs (IEPs). Knowledgeable nurses follow up with every patient by phone after visits to check on their ability to obtain medications and understand their education. These nurses also coordinated with a multidisciplinary team before each visit to ensure comprehensive care."

Table 3: Provider-identified Top 10 Barriers to Care
Addressing these barriers through the targeted efforts highlighted above is essential for improving the quality of care and outcomes for SCD patients.

Top 10 Barriers to Care Ranked from More to Less Important		
Rank	**Type of Barrier**	**Additional Details**
1	**Transportation Challenges for Patients**	Transportation challenges impede patients' access to necessary health care services.
2	**Availability and Access to Specialist Care**	The lack of available specialists and difficulty accessing specialized care were highlighted as major obstacles.
3	**Insurance Coverage and Related Issues**	Complications with insurance coverage, including referrals and provider networks, were identified as critical barriers.
4	**Affordability/Accessibility of Treatments and Screening Tests**	Financial constraints and limited access to essential treatments and tests were significant challenges for patients.
5	**Time Constraints for Both Providers and Patients**	Both health care providers and patients face time limitations, affecting the quality and continuity of care.
6	**Effectiveness of Tracking Systems or Electronic Health Records (EHR)**	Health care providers reported that many patients chose not to utilize the Electronic Medical Record (EMR) system for scheduling appointments and communicating with medical professionals. This has resulted in difficulties in monitoring and follow-up.
7	**Issues Related to Patient Follow-Up**	Problems with ensuring consistent patient follow-up were identified as notable barriers, with high no-show rates potentially related to transportation or insurance issues complicating continuous care and monitoring.
8	**Adequacy of SCD Training for Health Care Providers**	Insufficient training and education for health care providers regarding SCD management, particularly concerning the care of adult patients.
9	**Language/Cultural Barriers Impacting Care-Seeking Behavior**	Language differences and cultural barriers significantly impact care-seeking behavior, causing delays in seeking care due to biases and misunderstandings.
10	**Patient Compliance with Prescribed Treatments or Care Plans**	Ensuring patient adherence to prescribed treatments and care plans remains a persistent challenge, exacerbated by lapses in medication and a lack of trust in the health care system.

Population Estimates and Demographics for People with SCD in Illinois

Establishing Population Estimates
The exact number of people living with SCD in the United States and Illinois is unknown. Because exact numbers do not exist, this report primarily uses aggregate data from Medicaid recipients with SCD as the closest approximation for the population.

Medicaid Recipient Data
CMS estimates that 50% to 60% of people with SCD are insured through Medicaid.[2] According to CMS, Illinois has a higher than the national prevalence of Medicaid beneficiaries with SCD.[2]

Between January 1, 2016, and December 31, 2022, 6,278 Medicaid recipients claimed sickle cell disease. This likely represents about half of the Illinois population with SCD.

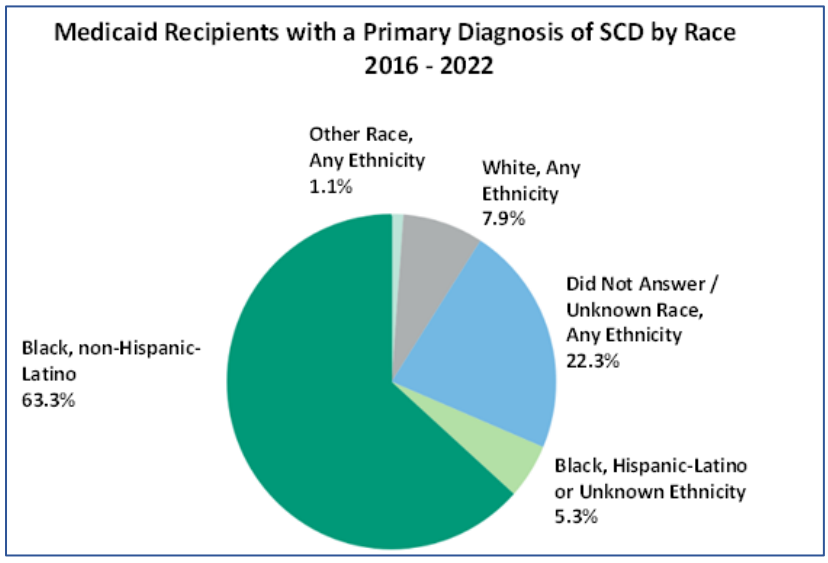
National Estimates
The Medicaid estimate is within range of what we would expect from applying national estimates. The Illinois populations at risk for SCD are 1.8 million Blacks/African Americans and 2.3 million Hispanics/Latinos.

- **We would expect approximately 5,000 people to be living in Illinois with sickle cell disease**.
  Based on the Centers for Disease Control and Prevention (CDC) national estimate of 1/365 Black /African American births and 1/16,300 Hispanic/Latino births having sickle cell disease, and the 2020 population of Illinois.

- **We would expect approximately 139,000 individuals with sickle cell trait to live in the state.**[4]
  Based on the CDC national estimate of 1/13 Black or African American births having sickle cell trait.

Figure 1: Medicaid Recipient Race Data

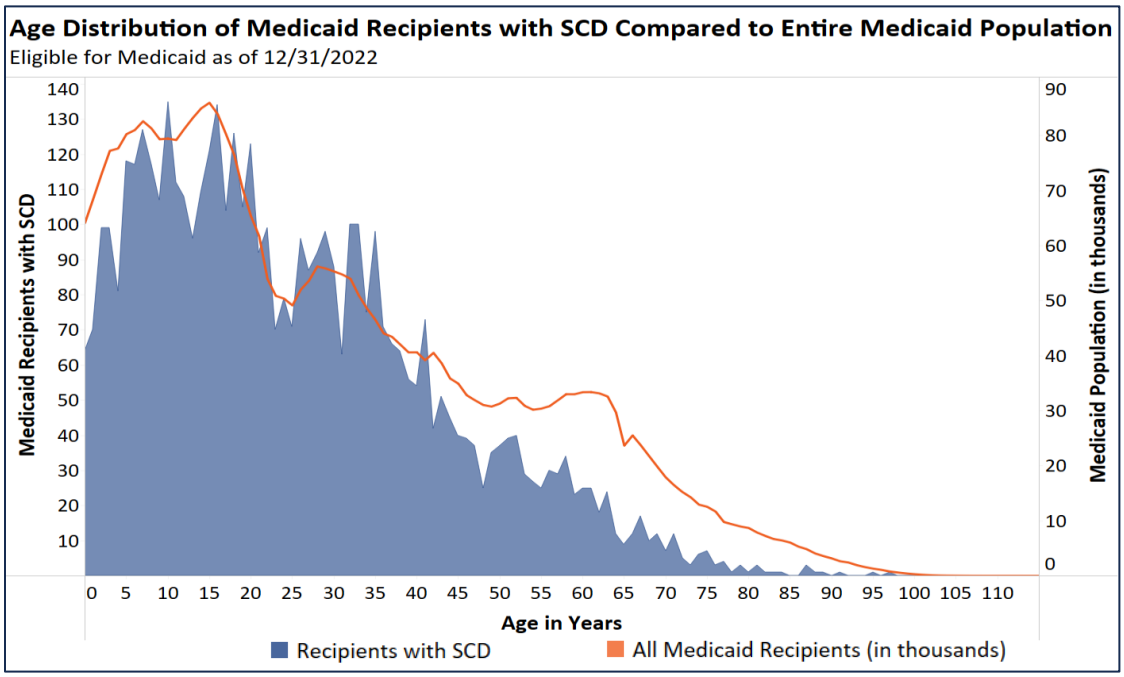
Most Medicaid recipients (63.3%) with an identified race are non-Hispanic Black. According to the CDC, more than 90% of people with SCD in the United States are non-Hispanic Black or African American, and an estimated 3%–9% are Hispanic or Latino.[1]



[4] Illinois Population by Race for Illinois and its Counties

Age Distribution for Medicaid Recipients Living with SCD

Figure 2: The age distribution graph below reflects that Medicaid recipients with SCD are not living as long as Medicaid recipients overall. There is a bump in Medicaid recipients during ages 50 to 65, whereas the number of recipients with SCD begins a steep decline at age 35.



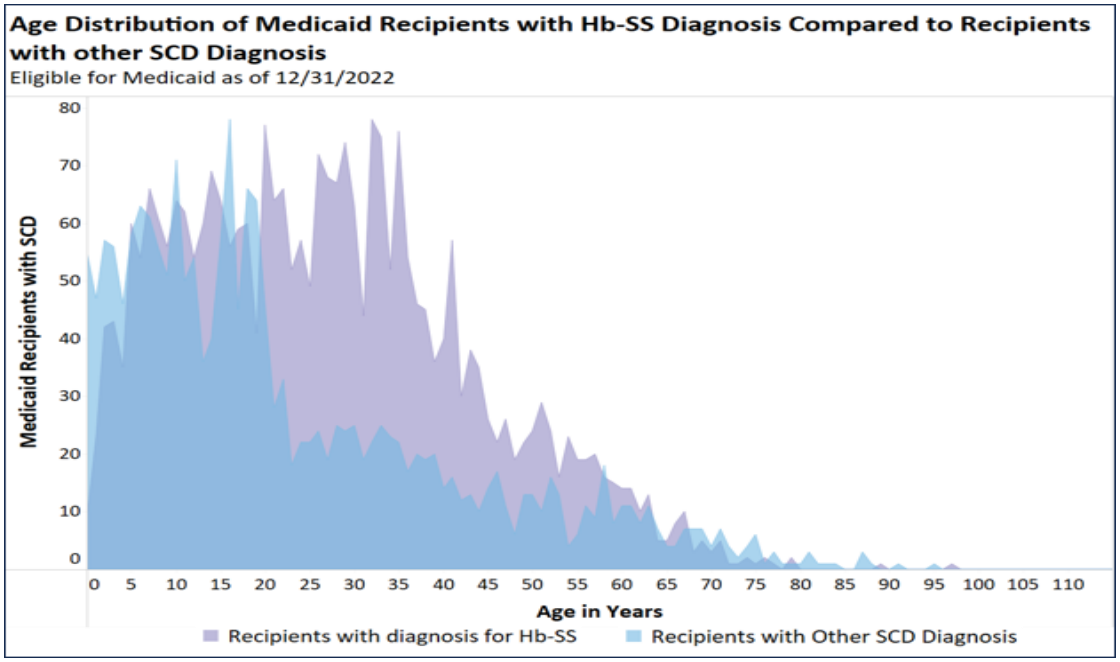


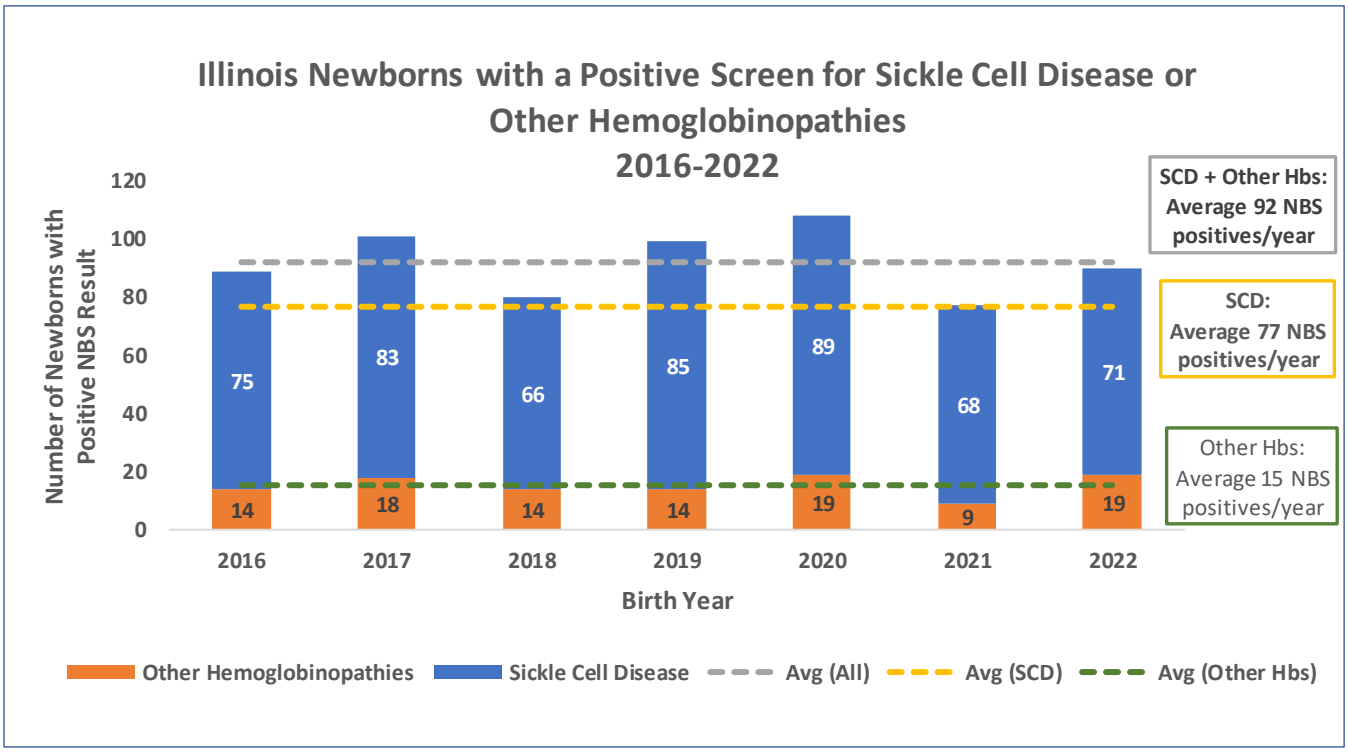
Figure 3: Reflecting the more serious nature of the disease, Medicaid recipients with an SCD diagnosis of Hb-SS, shown in purple above, are more likely to have Medicaid coverage as adults than SCD recipients with another subtype of SCD. The age distribution of SCD recipients who do not have Hb-SS more closely follows the overall distribution of Medicaid recipients.

IDPH Newborn Screening Data for Sickle Cell Disease and Other Hemoglobinopathies
An important public health program to prevent serious illness (Figure 4, Table 4)

Newborn screening helps babies with serious health conditions get off to a healthy start through early diagnosis and treatment initiation. During this period (2016-2022), approximately 969,000 Illinois babies were screened for sickle cell disease and other hemoglobinopathies. Newborns with a positive screen, suggesting a risk for disease or trait, were referred for clinical follow-up and confirmation of the diagnosis. Differences in newborn screening rates across years may be due to natural chance, population shifts, and changing birth rates.

Summary

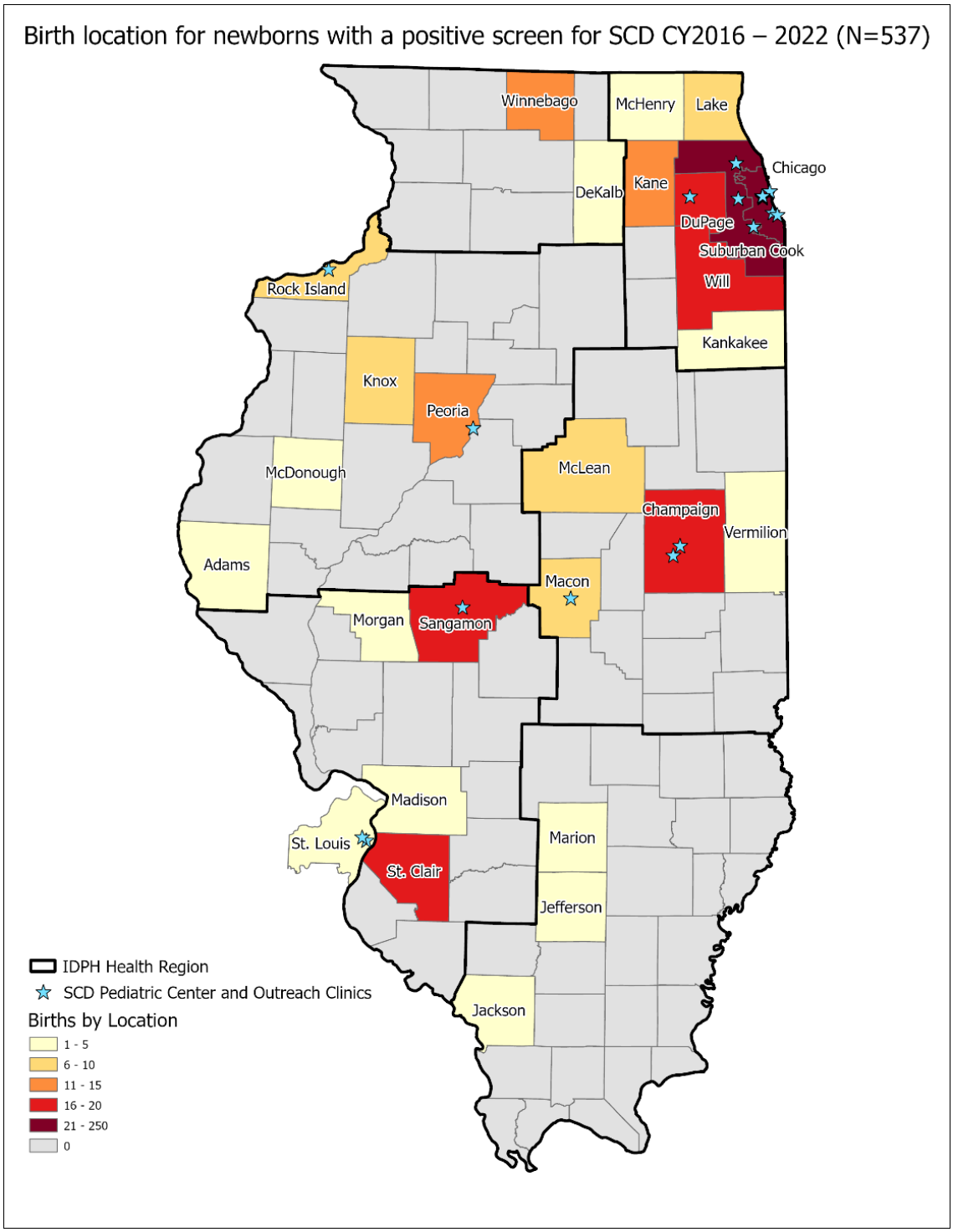
- In total, 644 newborns had a positive screen for SCD or other hemoglobinopathy.
- A positive screen for SCD (83%) was more common than for other hemoglobinopathies.
- Most (51%) screened positive for the more serious form of SCD (sickle cell anemia).
- An estimated 39,900 had newborn screening results showing a high likelihood of sickle cell trait.



Newborns with a Positive Screen for Sickle Cell Disease & Other Hemoglobinopathies		
Group	**Subtype**	**Number of newborns with positive NBS result (%)**
Sickle Cell Disease (SCD)	Sickle cell anemia	331 (51%)
	Sickle hemoglobin C disease	160 (25%)
	Sickle beta thalassemia disease	46 (7%)
Other Hemoglobinopathies (Hbs)	Non-sickling hemoglobinopathies	107 (17%)
Total		**644**

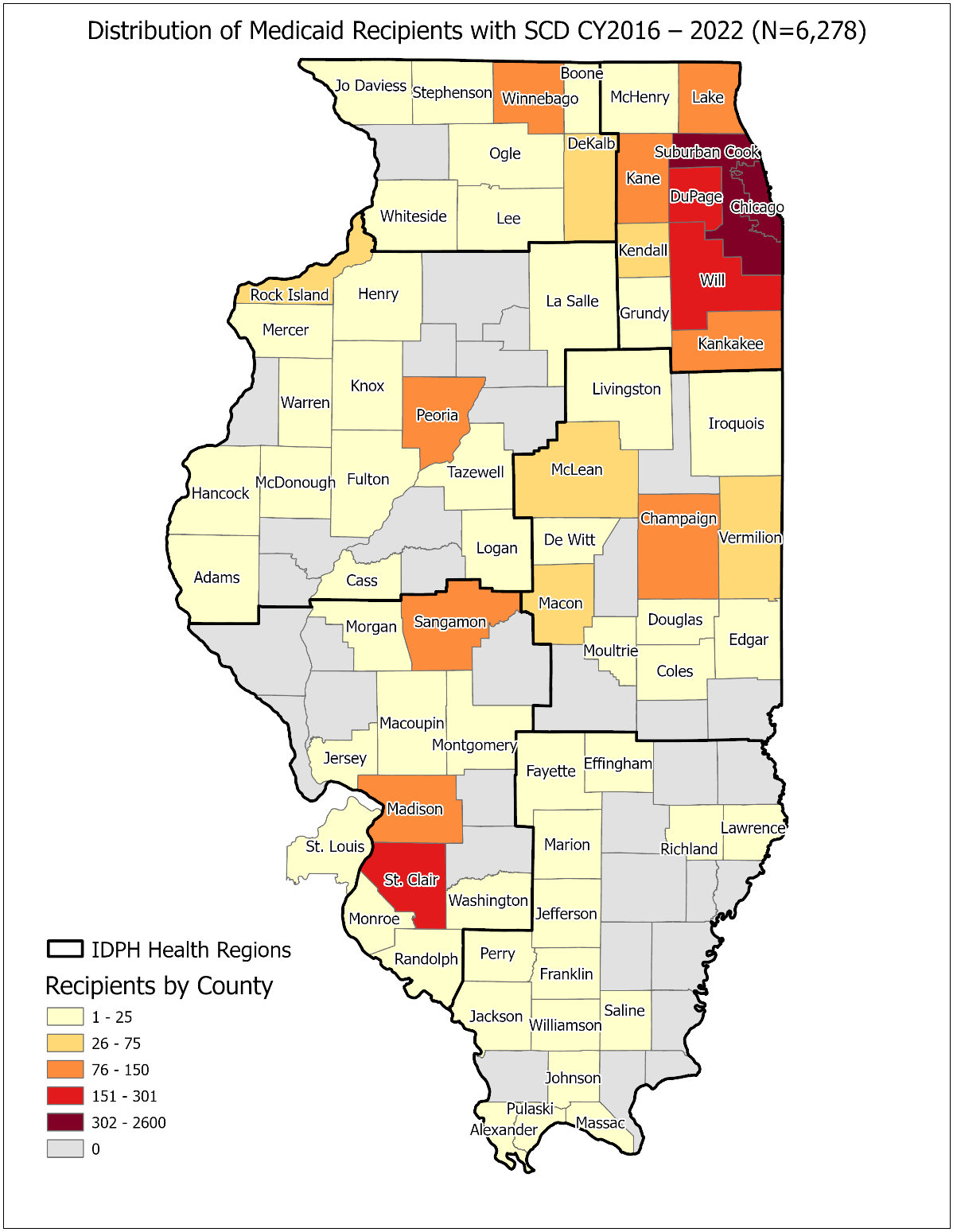
Geographic Location of Illinois residents with SCD

Map 1: Birth Location Roughly Reflects Population Centers for People with SCD
The map includes hospitals, birth centers, and home births. Also shown are the SCD pediatric centers and outreach clinics where newborns with a positive screen for SCD or sickle cell trait are referred for confirmation and evaluation. A list of these centers and outreach clinics is included in Appendix B.



Map 2: Distribution of Medicaid Recipients with an SCD Primary Diagnosis 2016 – 2022

The map shows the distribution **of 6,278 Medicaid** recipients with SCD aggregated by county. If recipients had more than one address, the most recent address was used. Most of these individuals live in the Chicago metropolitan area.



Health Care Access for Persons with SCD in Illinois

Tables 5 and 6: Medicaid Sickle Cell Disease Diagnosis Data
Most Medicaid recipients with an SCD subtype identified had sickle cell anemia (Hb-SS) and were more likely to have severe complications.

Medicaid Primary Diagnosis Claims with Sickle Cell Subtype Identified 1/1/16 and 12/31/22			
ICD-10 Code SCD Categories	**Subtype**	**# of Medicaid Recipients (%)**	**Claims**
D57.0	Hb-SS disease	3,788 (83%)	437,017 (85%)
D57.2	Sickle-cell/Hb-C	587 (13%)	50,751 (10%)
D57.4	Sickle-cell thalassemia	19 (4%)	28,035 (5%)

Medicaid Claims for Severe Complications of Sickle Cell Disease 1/1/16 and 12/31/22	
Diagnoses (Complications of SCD)	**Number of Claims**
SCD Crisis	455,408
Acute Chest Syndrome	33,282
Splenic Sequestration	1,892
Cerebral Vascular Involvement	1,517
Crisis with Other Complication	3,289

Description of SCD Severe Complications (Source: CDC)
Severe complications of SCD are common, and more likely to occur in people who have sickle cell anemia (the Hb-SS form of the disease).

- **SCD Crisis:** A defining feature of SCD occurs when sickled cells block blood flow to an area or organ of the body, causing severe pain and organ damage. A crisis can be brought on by high altitudes, dehydration, illness, stress, or temperature changes.

- **Acute Chest Syndrome (ACS):** ACS is a life-threatening complication that can result in lung injury, breathing difficulty, and low oxygen to the rest of the body. ACS may occur when sickled cells block blood and oxygen from reaching the lungs or may be caused by a viral or bacterial infection.

- **Splenic Sequestration:** When sickled red blood cells are trapped in the spleen, the rest of the body does not get enough oxygen. If not treated, an acute spleen can cause the body to go into shock. It is an emergency and may be life-threatening.

- **Cerebral Vascular Involvement (Stroke):** Stroke is a catastrophic complication of SCD and a leading cause of death in both children and adults.

- **Crisis with Other Complication:** This refers to a situation in which an acute crisis occurs in combination with one of the other severe complications.

Map 3: Health Care Provider Location and Number of Medicaid Claims

The map shows the number of claims from providers who cared for Medicaid recipients with a primary diagnosis of SCD during 2016 -2022. The highest number of claims was in the Chicago metropolitan area. In most counties, providers are giving some care to Medicaid recipients with SCD, and some care was provided in neighboring states. The types of providers giving care are listed in the table on the next page.

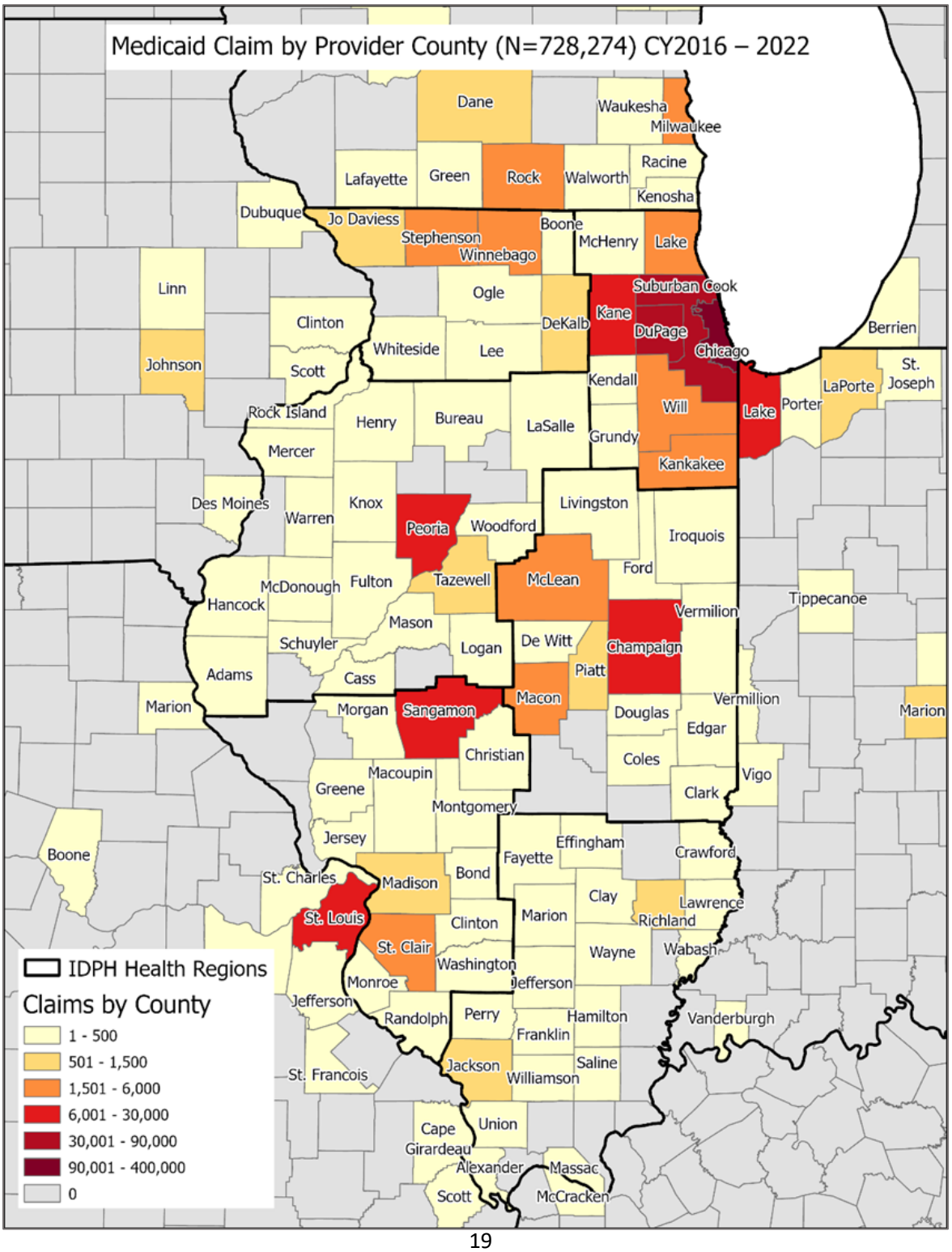


Table 7: Health Care Provider Type for Medicaid SCD Claims (mapped above)
Physicians and hospitals accounted for the majority (94%) of Medicaid claims for individuals with an SCD primary diagnosis during this period.

Medicaid Claims by Provider Type for SCD Primary Diagnosis 1/1/16 and 12/31/22 (Excludes Pharmacy*)	
Provider Type	**Claims**
Physicians (both inpatient and outpatient)	543,120
General Hospitals	142,472
Nurse Practitioners	22,470
Independent Laboratories	5,240
Transportation Services	4,289
Other Providers of Medical Equipment/Supplies (Non-registered)	3,216
Physician Assistant Only	2,633
Federally Qualified Health Centers (FQHC)	1,609
Home Infusion and Other Surgical Supplies	899
Encounter Rate Clinic (not an FQHC or RHC)	555
Nursing Facilities	401
Home Health Agencies - In Home	393
Optometrists	270
School-based Health Centers	172
Other Provider Type †	149
Waiver Service Provider -- Disability (DHS/DRS)	147
Waiver Service Provider -- Elderly (DOA)	143
Psychologist	94
Rural Health Clinics (RHC)	90
Total Claims	**728,362**

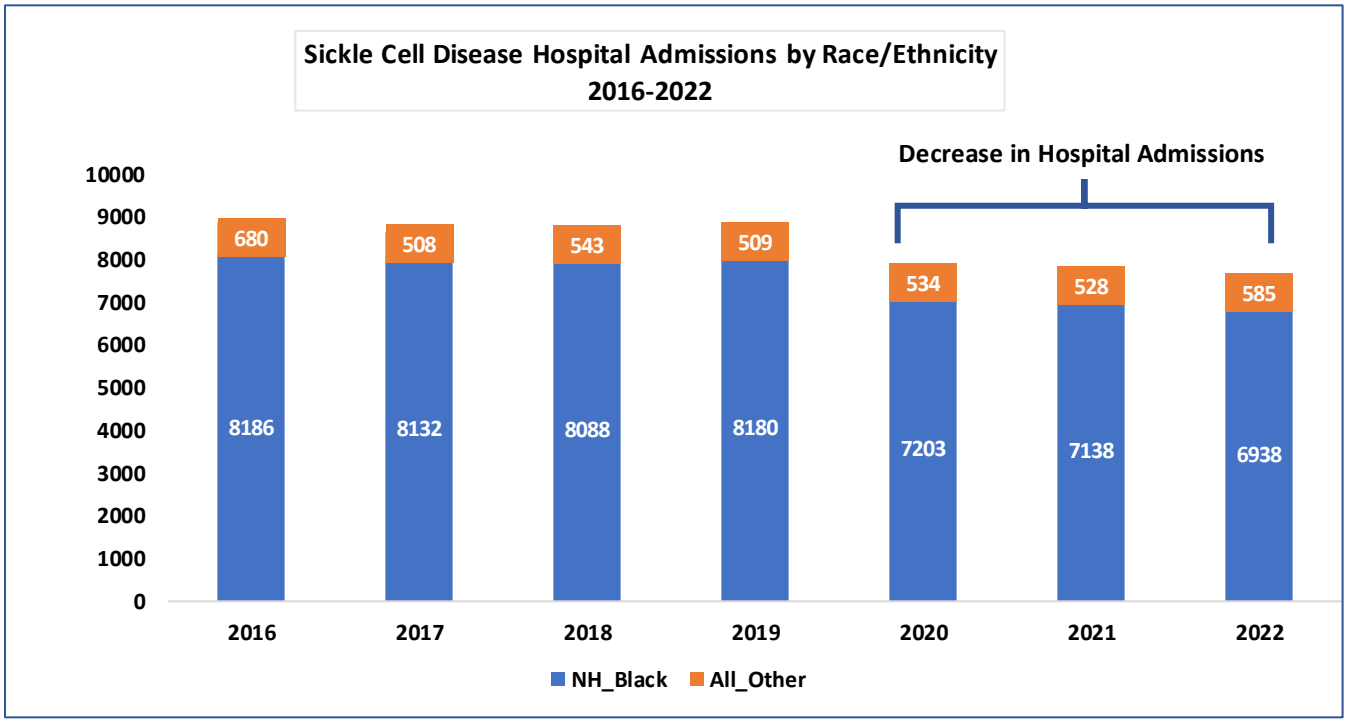
Notes:
*Medications dispensed in pharmacies or administered by non-institutional providers are not associated with diagnosis codes. This table does not include these 530,622 claims over the entire period.
†"Other provider types" include a wide range of entities, such as ambulatory surgical treatment centers, hospice, physical therapists, and clinical social workers.

IDPH Illinois Hospital Utilization Data for People with SCD (Regardless of Payor)
This data is from Illinois hospitals only. Care for Illinois residents is also provided at hospitals in neighboring states, most notably in St. Louis, Missouri.

- **Emergency Department Visits (resulted in a release after treatment):** Between 2016 and 2022 over **62,500** emergency department visits related to sickle cell disease were recorded in Illinois. This represents an average of more than 8,900 emergency department visits a year. Individuals living with SCD have high levels of complications from their disease, resulting in frequent utilization of the emergency department. Most commonly these visits are to control pain.[5]

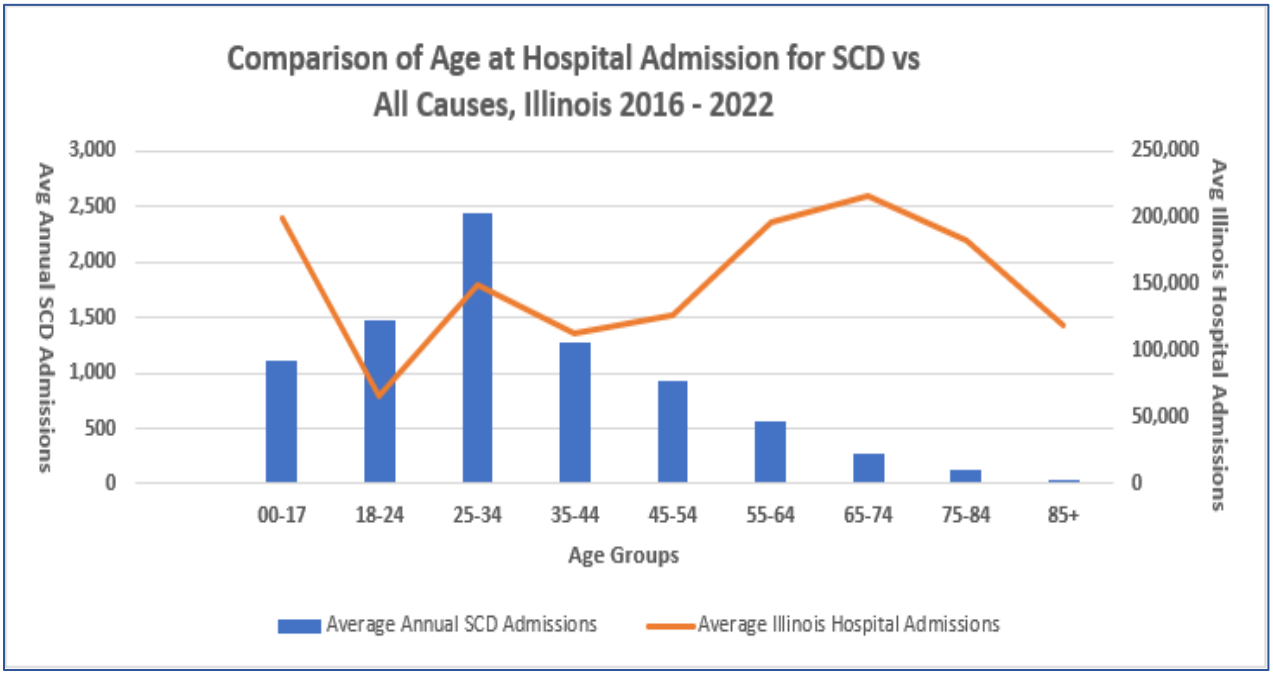
- **Hospital Admissions:** Between 2016 and 2022, 57,752 hospital admissions related to sickle cell disease were recorded in Illinois. This represents an average of over 8,250 admissions a year.

Race/Ethnicity (Figure 5)
Most people with SCD in Illinois are non-Hispanic Blacks, and this is reflected in the race and ethnicity data for hospital admissions shown in the table on the next page. Of note, there was an approximately 12% decrease in hospital admissions during 2020-2022. Additional evaluation is needed to determine if the decrease was related to less access to care during the COVID-19 pandemic or another cause.



[5] Attell BK, Barrett PM, Pace BS, et al. Characteristics of Emergency Department Visits Made by Individuals with Sickle cell Disease in the U.S., 1999-2020. *AJPM Focus 2024;3(1):100158.* https://doi.org/10.1016/j.focus.2023.100158

Figure 6: Age at Hospitalization – Illinois Data is Consistent with National Trends
Hospital admissions for patients with SCD peak in early to middle adulthood when progressive organ damage from the disease begins to become apparent.[6] This contrasts with the overall Illinois population where hospital admissions peak at age 65 years and beyond, typically for end-of-life care. Most people with SCD do not reach 65 years of age. (Note: the high point for 00-17 overall is due to hospital births.)

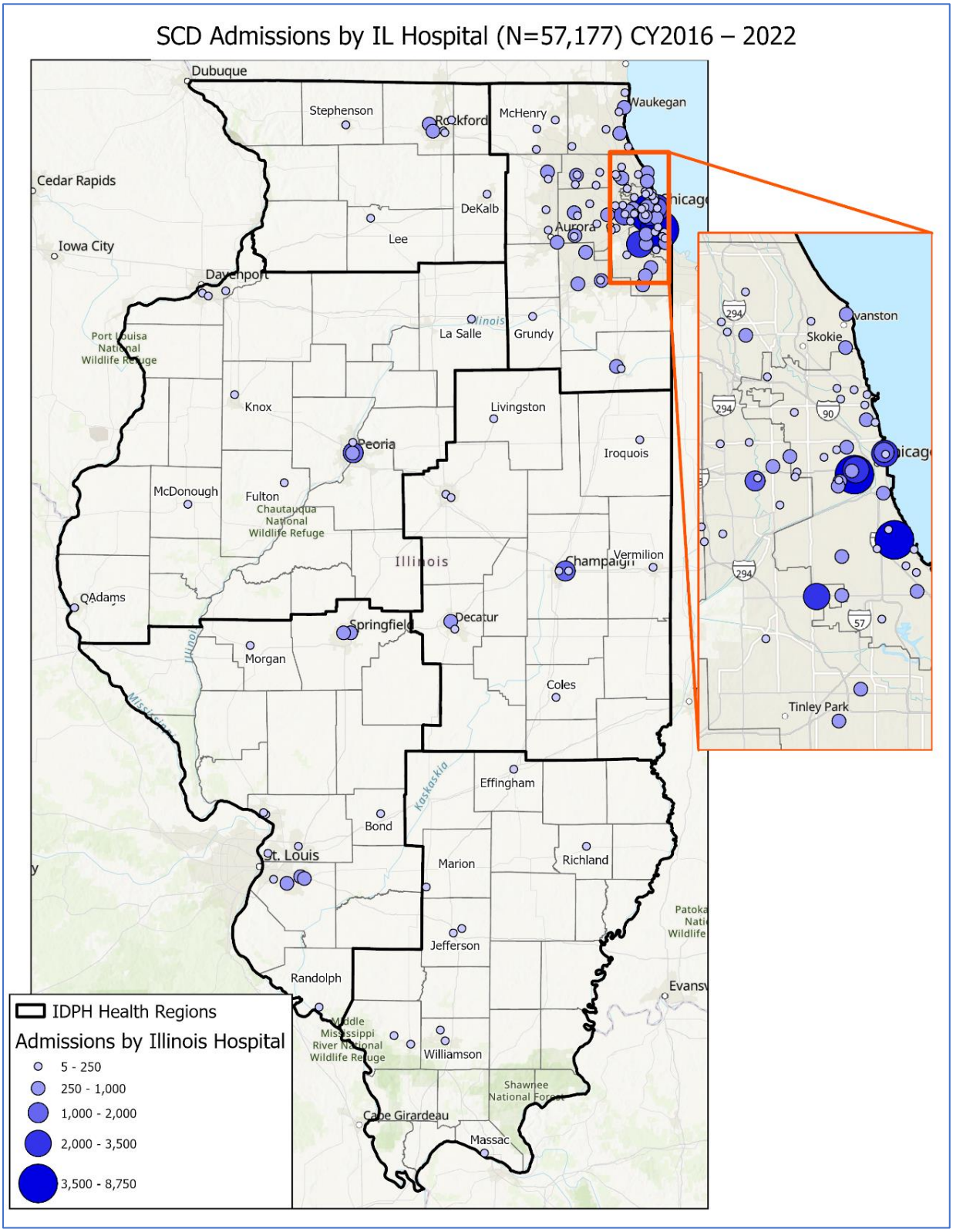


Ranking of Illinois Hospitals by Number of Admissions for People with SCD (Table 8)
Most people with SCD were admitted to hospitals located in Chicago or suburban Cook County, with the University of Illinois and the University of Chicago handling about half of all admissions.

Top 10 Illinois Hospitals for Admissions of Patients with SCD 2016 - 2022			
Rank	**Hospital Name**	**Admissions**	**City**
1	UI Health	8,663	Chicago
2	UChicago Medicine	8,091	Chicago
3	Northwestern Memorial Hospital	3,448	Chicago
4	Rush University Medical Center	2,962	Chicago
5	Advocate Christ Medical Center	2,709	Oak Lawn
6	Ann & Robert H. Lurie Children's Hospital of Chicago	1,856	Chicago
7	Loyola University Medical Center	1,451	Maywood
8	Carle Foundation Hospital	1,418	Urbana
9	OSF Saint Francis Medical Center	1,328	Peoria
10	John H. Stroger Jr. Hospital of Cook County	867	Chicago

[6] Chennapragada SS, Thevuthasan S, Savani S, et al. Characteristics and Trends of Hospitalizations for Sickle-Cell Related Complications. *Blood (2023) 142 (Supplement 1): 2305*. https://doi.org/10.1182/blood-2023-187511

Map 4: SCD Admissions by Illinois Hospital (Source: IDPH Hospital Admission Data)
Hospital care was concentrated in the Chicago metropolitan area. However, people with SCD also receive hospital care outside of Illinois, particularly at hospitals in St. Louis, Missouri.



Illinois Mortality Data by Race/Ethnicity, Sex, and Age for People with SCD

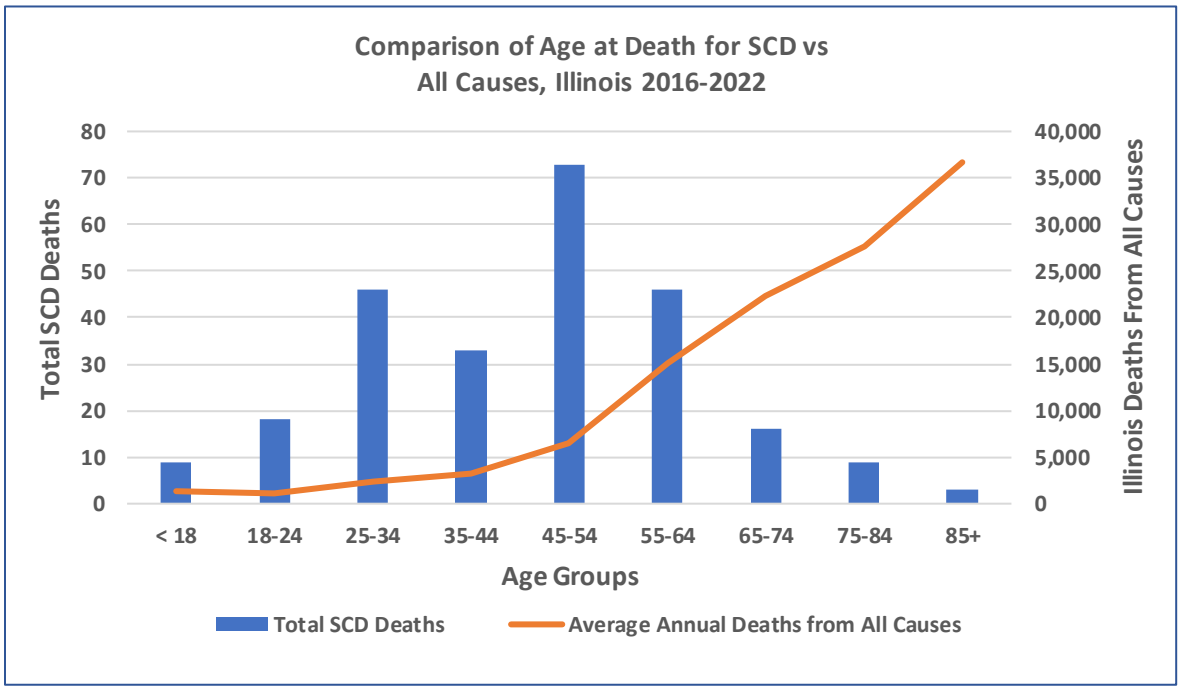
Mortality by Sex and Race (Table 9)
The table shows all deaths during this seven-year period in which "sickle-cell disorder" was listed on the death certificate as an "underlying" or "contributing cause" of death.

- 96% of deaths were in non-Hispanic Blacks.
- Men and women were equally likely to die from sickle cell disease.

Differences in Life Expectancy (Figure 7)
People with sickle cell disease in Illinois have a life expectancy much less than the overall population, typically dying before age 65 years. This is consistent with national data showing the average life expectancy for someone living with SCD to be 52.6 years[7].

Illinois Sickle Cell Disease Deaths by Race and Sex 2016-2022

		Sex		Race/Ethnicity	
Year	Total	Male	Female	NH Black	Other
2016	38	24	14	36	2
2017	31	13	18	31	0
2018	28	14	14	27	1
2019	38	19	19	36	2
2020	45	23	22	42	3
2021	34	13	21	34	0
2022	39	21	18	36	3
Total	**253**	**127**	**126**	**242**	**11**
Ave.	**36**	**18**	**18**	**35**	**2**



[7] Jiao B, Johnson K, Ramsey S, Bender M, Devine B, Basu A. Long-term survival with sickle cell disease: a nationwide cohort study of Medicare and Medicaid beneficiaries [published online, 2023 Mar 16]. *Blood Adv*. doi: 10.1182/bloodadvances.2022009202.

Establishing an Illinois Sickle Cell Impact Surveillance System (ISCISS) What Was Accomplished and Planned Next Steps

This first "Sickle Cell Statewide Impact and Surveillance Program" report is an initial step towards developing an understanding of the challenges people who live with SCD have with accessing needed health care. The significant negative impact that the disease can have on individuals is clear from the Illinois data, and a call for action. IDPH will continue to work with HFS and SCD health care providers to improve access to care and outcomes for people with SCD. A priority will be to engage with people who have SCD and advocacy groups, such as the Sickle Cell Disease Association of Illinois (SCDAI).

The goal of the sickle cell surveillance system laid out by Public Act 102-004 was to determine:

1. The prevalence, by geographic location, of people diagnosed with sickle cell disease and sickle cell trait in Illinois.
2. The availability and affordability of screening services for sickle cell trait in Illinois.
3. The location and capacity for the treatment of sickle cell disease and sickle cell trait.
4. The unmet medical, psychological, and social needs of people in Illinois with sickle cell disease.
5. The underserved areas of Illinois for the treatment of sickle cell disease.

How these questions were addressed in this report
The overarching concern is to evaluate whether people with SCD can get the care that they need, to identify barriers, and to develop a plan to eliminate or minimize barriers. Following the summary for each area are the planned next steps. The primary means of IDPH addressing these issues is through grants from the "Sickle Cell Disease Prevention, Care, and Treatment Program" to entities that provide care and services to affected individuals. (See the Acknowledgement Section on page 5).

1. **Prevalence by Geographic Location**
   Initial estimates for the size and geographic location of the population included in this report were made using birth location for newborns with a positive screen and Medicaid recipient claims data. Based on this information, most people in Illinois with SCD live in the Chicago metropolitan area, with a substantial number also living in St. Clair and Madison counties.

   Next Steps
   - IDPH will hire dedicated staff to continue the work begun with this report of pulling together diverse data sources and collaborating with other agencies to better understand the affected population, health care access barriers, and unmet needs.
   - Linking data from multiple IDPH programs will streamline data analysis, allow for a more in-depth assessment of trends, and give more robust results.
   - Through discussions with health care providers, community organizations, and people living with SCD, IDPH will assess the accuracy of the population estimates included in this report.

2. **Availability and Affordability of Screening Services**
   Newborns in Illinois are screened for SCD shortly after birth. The IDPH Newborn Screening Program ensures that newborns with a positive screen are seen by a specialist to confirm the diagnosis, be evaluated, and

receive further care. Newborn screening is a mandated public health service. The IDPH laboratory does not provide SCD clinical confirmatory testing or testing for adults. These tests must be obtained through a health care provider.

For many reasons, people with sickle cell trait may not know their trait status. Parents of babies diagnosed with SCD, prospective parents with a family history of sickle cell disease, and people more likely to have sickle cell trait due to their ancestry, may seek clinical screening for sickle cell disease.

Next steps

- IDPH has received anecdotal reports that there may be barriers to adults accessing SCD carrier screening and counseling. A follow-up assessment will be made to determine the accessibility and affordability of these services.

3. **The location and capacity for the treatment of sickle cell disease and sickle cell trait.**
For this first report, Medicaid claims data were used to map the location where care was provided and the number of claims by location to get an initial impression of how care was distributed across the state. Hospital admission data was similarly mapped. The majority of care is being provided in the Chicago metropolitan area, with health care professionals in Champaign, Peoria, Springfield, and St. Louis also providing substantial levels of care.

Next steps

- Interview health care providers specializing in SCD, community-based organization staff, and people living with SCD to identify areas of the state where capacity is insufficient to meet needs. This will include access to infusion centers, pain management therapies, and other recommended preventive services and treatments.
- Conversations with SCD health care providers will center on national guidelines for SCD care.
- Assess Medicaid Managed Care Organization (MCO) SCD services, focusing on standards of care as well as specific outreach and care management initiatives for Medicaid recipients in Illinois.

4. **The unmet medical, psychological, and social needs of people in Illinois with sickle cell disease.**
A survey of providers was conducted, and the top three barriers to obtaining care were identified as transportation challenges for patients, availability of and access to specialist care, and insurance coverage issues. In addition, a review of Illinois hospital admissions and emergency department visit data, Medicaid claims, and mortality data has revealed that people with SCD have severe complications from their disease requiring frequent hospital admissions from early adulthood and are dying at a much younger age than the Illinois population.

Next steps

- Have listening sessions with health care providers, community-based organization staff, and people living with SCD, to develop a deeper understanding of the multiple identified barriers to receiving needed care. This will include social and mental health services.
- Assess how well youth with SCD are transitioning from pediatric to adult health care. Becoming uninsured or having uncoordinated care is a risk for this age group.

5. **The underserved areas of Illinois for the treatment of sickle cell disease.**
   This first assessment and report utilized multiple data sources to determine which areas of the state have population centers for people with SCD and where they receive care. At the geographic level, most people with SCD live in areas where care is available; however, whether they can easily access that care is a question that requires further investigation. Some SCD care is being provided in most areas of the state, as well as in neighboring states.

   Next steps
   - IDPH staff have received anecdotal reports suggesting greater barriers to accessing care for people with SCD who reside outside of those areas where care is available. Future work will include assessments of provider type, distance from care, transportation challenges, and other issues identified during listening sessions or other channels.
   - Assess the need to provide training in SCD care for primary care providers, to increase the capacity for caring for people with SCD, particularly in areas where specialist care may not be available.

Appendix A: Summary of Survey Questions for Providers

Section 1: Provider's Information

- What type of facility do you work in or collaborate with for SCD care?
- What is your role within your organization?
- How many years have you been working with SCD patients?
- In which of the IDPH Public Health Regions do you primarily provide services for SCD patients?
- Can you provide an estimate of the number of SCD patients in your practice?
- What is the age range of the SCD patients in your practice?
- Describe the range of services your facility offers to SCD patients.

Section 2: Barriers and Challenges

- What barriers to effective SCD care have you encountered?
- Please provide more detail on your top three (or more) selected barriers, their impact, and any strategies you've employed to overcome them.

Section 3: Success Stories and Effective Strategies

- Can you share any success stories or effective strategies that have significantly improved care or outcomes for patients with SCD in your practice?
- Have there been any notable collaborations with the community or other organizations?
- Can you briefly describe any notable collaborations with the community or other organizations?

Section 4: Data Collection and Reporting

- What data do you believe are most important to include in an IDPH SCD Impact Report?
- Do know any other organizations, agencies, provider types, service providers, or others who may have important information to share?
- Do you have any additional insights or comments regarding your experience in providing care for SCD patients or any suggestions on how care for SCD patients can be improved?

Appendix B: List of SCD Pediatric Centers and Outreach Clinics

Clinic Name / Facility	City	State
La Rabida Children's Hospital	Chicago	Illinois
Ann and Robert H. Lurie Children's Hospital of Chicago	Chicago	Illinois
St. Louis Children's Hospital/Washington University School of Medicine	Saint Louis	Missouri
OSF HealthCare Children's Hospital of Illinois	Peoria	Illinois
The University of Illinois Hospital and Health Sciences System (UIC)	Chicago	Illinois
Keyser Center at Advocate Children's (Advocate Christ, Advocate Hope)	Oak Lawn	Illinois
John H. Stroger Jr. Hospital	Chicago	Illinois
Rush University Medical Center	Chicago	Illinois
Advocate Medical Centers Lutheran General Children's Medical Center	Park Ridge	Illinois
Edward Hospital- Naperville (UC/ Comer Children's Hospital)	Chicago	Illinois
St. Jude's Midwest Affiliate Clinic	Peoria	Illinois
Rock Island County Health Department	Rock Island	Illinois
Macon County Health Department	Decatur	Illinois
Loyola University Medical Center	Maywood	Illinois
Central DuPage Hospital	Winfield	Illinois
SIU Healthcare/ St. John's Children's Hospital	Springfield	Illinois
Carle Clinic	Champaign	Illinois
Cardinal Glennon Children's Medical Center (Bob Costas Center)	Saint Louis	Missouri
Milwaukee Campus - Children's Wisconsin	Milwaukee	Wisconsin
Hektoen Institute	Chicago	Illinois
OSF Heart Of Mary Medical Center	Urbana	Illinois

APPENDIX C – List of ICD-10 Codes Used to Find Cases of Sickle Cell Disease

ICD-10 Code	Diagnosis Description
D57.00	Hb-SS disease with crisis, unspecified
D57.01	Hb-SS disease with acute chest syndrome
D57.02	Hb-SS disease with splenic sequestration
D57.03	Hb-SS disease with cerebral vascular involvement
D57.09	Hb-SS disease with crisis with other specified complication
D57.1	Sickle-cell disease without crisis
D57.20	Sickle-cell/Hb-C disease without crisis
D57.211	Sickle-cell/Hb-C disease with acute chest syndrome
D57.212	Sickle-cell/Hb-C disease with splenic sequestration
D57.213	Sickle-cell/Hb-C disease with cerebral vascular involvement
D57.218	Sickle-cell/Hb-C disease with crisis with oth complication
D57.219	Sickle-cell/Hb-C disease with crisis, unspecified
D57.40	Sickle-cell thalassemia without crisis
D57.411	Sickle-cell thalassemia, unsp, with acute chest syndrome
D57.412	Sickle-cell thalassemia, unsp, with splenic sequestration
D57.413	Sickle-cell thalassemia, unsp, with cerebral vascular invl
D57.418	Sickle-cell thalassemia, unsp, with crisis with oth comp
D57.419	Sickle-cell thalassemia, unspecified, with crisis
D57.42	Sickle-cell thalassemia beta zero without crisis
D57.431	Sickle-cell thalassemia beta zero with acute chest syndrome
D57.432	Sickle-cell thalassemia beta zero with splenic sequestration
D57.438	Sickle-cell thalassemia beta zero with crisis with oth comp
D57.439	Sickle-cell thalassemia beta zero with crisis, unspecified
D57.44	Sickle-cell thalassemia beta plus without crisis
D57.451	Sickle-cell thalassemia beta plus with acute chest syndrome
D57.452	Sickle-cell thalassemia beta plus with splenic sequestration
D57.458	Sickle-cell thalassemia beta plus with crisis with other comp
D57.459	Sickle-cell thalassemia beta plus with crisis, unspecified
D57.80	Other sickle-cell disorders without crisis
D57.811	Other sickle-cell disorders with acute chest syndrome
D57.812	Other sickle-cell disorders with splenic sequestration
D57.813	Other sickle-cell disorders with cerebral vascular invl
D57.818	Other sickle-cell disorders with crisis with oth comp
D57.819	Other sickle-cell disorders with crisis, unspecified

Note: Excludes D57.3, Sickle Cell Trait