



Sickle Cell Disease  
Newborn Screening Surveillance Report

October 2014

*Celebrating*  
**27**  
*Years*

of Sickle Cell Screening

**Michigan Department of Community Health**

Lifecourse Epidemiology and Genomics Division  
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**Learn more about Michigan Hemoglobinopathy Surveillance:**

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## ***Executive Summary***

Staff from the Michigan Department of Community Health (MDCH), the Sickle Cell Disease Association of America—Michigan Chapter (SCDAA-MI), and members of the Michigan Hemoglobinopathy Quality Improvement Committee (MiHemQIC) collaborated to assess the burden of hemoglobin disorders on individuals of all ages identified primarily through Michigan’s Newborn Screening (NBS) Program. From 2009-2013, systematic data collection 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 was a priority in Michigan. Subsequently, key baseline measurements relevant to important aspects of sickle cell disease (SCD) in Michigan were developed.

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, emergency department visits, inpatient hospitalizations, and outpatient visits. These data span various time periods and were obtained from birth certificate records, the SCDAA-MI’s records describing children and adults served through the center, immunization information from the Michigan Care Improvement Registry (MCIR), and Medicaid and Children’s Special Health Care Services (CSHCS) claims from Michigan’s data warehouse.

This report describes the twelve hemoglobinopathy indicators for Michigan, as well as comments about their significance and potential utility. Michigan data on these twelve indicators provide benchmarks for hemoglobinopathies related to Healthy People 2020 objectives. Further, the findings provide important information to identify gaps in services, and represent an essential step for planning public health action.

From 1988-2011, a total of 1,689 newborns were detected and confirmed with SCD in Michigan through the NBS Program; 95 percent of newborns detected were black. In 2011, 1 in 364 screened African-American newborns and 1 in 1,907 screened newborns were diagnosed with SCD; 61 newborns total were diagnosed with SCD. An additional 2,817 newborns were identified as having sickle cell trait (SCT) on initial screening results in 2011. *Other notable findings for children and youth with SCD include:*

- 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%).
- Based on Michigan Medicaid programs claim 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 had a claim for transcranial doppler screening.
- The percent of children with four or more claims for emergency department visits increased from 16 percent in 2008 to 19 percent in 2011.

## ***Acknowledgments***

This report is the product of the collaborative efforts of many people at the Michigan Department of Community Health, as well as critical support from partner agencies including the Sickle Cell Disease Association of America - Michigan Chapter (SCDAA-MI). We thank the other members of the Registry and Surveillance System for Hemoglobinopathies (RuSH) team at the Centers for Disease Control & Prevention (CDC) for their review and commentary and also thank the Michigan Hemoglobinopathy Quality Improvement Committee (MiHemQIC) for their input and contributions. *Contributing members of the MDCH team include:*

- Janice Bach, State Genetics Coordinator and Genomics & Genetics Manager
- Erika Garcia, Medicaid Data Analyst
- Mary Kleyn, Newborn Screening Epidemiologist
- Sarah Lyon-Callo, Lifecourse Epidemiology and Genomics Division Director
- Dominic Smith, Hemoglobinopathy Program Coordinator
- Robin Stottlemeyer, Newborn Special Projects Coordinator
- William Young, Newborn Screening Follow-up Manager

This work was supported by the Hemoglobinopathy Surveillance Quality Improvement Program (MiHemSQIP), as part of Michigan's RuSH pilot project. RuSH was funded by the National Heart, Lung, and Blood Institute through a cooperative agreement with the CDC.

## Background

Sickle cell disease (SCD), often called “sickle cell anemia,” is the most commonly inherited blood disorder in the United States.<sup>1</sup> This condition causes red blood cells to form an abnormal crescent shape that is rigid and sticky which leads to clogged or closed blood vessels. Persons living with SCD have life-long complications such as stroke, splenic sequestration, acute chest syndrome, vision loss, and pulmonary embolism. In addition, individuals can frequently experience severe pain episodes know as “crises.” SCD is associated with premature mortality.

A national policy was first recommended by a 1987 National Institutes of Health Consensus Conference panel following the publication of a randomized controlled trial that demonstrated a 84 percent reduction in the incidence of infections in children with SCD who received prophylactic oral penicillin starting at four months of age, compared to children taking placebo.<sup>2</sup> Affected newborns will appear normal, but anemia develops in the first few months of life, followed by increased susceptibility to infection, slow growth rates and the possibility of life threatening splenic sequestration.<sup>3</sup> With appropriate medical care including penicillin prophylaxis, appropriate vaccinations, and long term management, the complications of SCD can be minimized.

Newborn screening (NBS) for SCD has been conducted at the Michigan Department of Community Health since 1987. Longitudinal follow-up of SCD case patients diagnosed by NBS traditionally has been carried out through five years of age by NBS follow-up staff in collaboration with the Sickle Cell Disease Association of America, Michigan Chapter (SCDAA-MI). SCDAA-MI is a community-based organization whose contract is funded through Michigan’s NBS fee to provide medical management and support to families of children in Michigan with SCD.

As part of the Centers for Disease Control & Prevention (CDC) —funded Registry and Surveillance System for Hemoglobinopathies (RuSH), the Hemoglobinopathy Surveillance Quality Improvement Program (MiHemSQIP) expanded hemoglobinopathy follow-up throughout the lifespan by enhanced monitoring of comorbidities, service use, and patient outcomes. The MiHemSQIP served as a Michigan-specific data source for hemoglobinopathies from 2010 to 2013. The dissemination of MiHemSQIP data is an essential step in translating findings into public health action. We are pleased to present this cumulative hemoglobinopathy surveillance report based on a compilation of findings from multiple data sources including birth certificate records, SCDAA-MI records, immunization information, NBS and Medicaid records, and claims from Medicaid and Children’s Special Health Care Services records.

MiHemSQIP aimed to assess the hemoglobinopathy burden in Michigan through ongoing surveillance. This current report provides information on hemoglobinopathy cases of all ages throughout Michigan, but highlights those identified through the NBS. It provides benchmarks for twelve hemoglobinopathy baseline measures, which can be examined in relation to *Healthy People 2020* objectives where applicable. With these findings, gaps in services may be identified and described in order to better understand and address the needs of the hemoglobinopathy population in Michigan. **Surveillance objectives include:**

- Monitor short-term patient follow-up indicators (i.e. antibiotic prophylaxis, confirmatory diagnosis, patient education) over time.
- Measure long-term trends in quality of care and outcomes specific to the hemoglobinopathy population of Michigan.
- Create benchmarks for comparison with national standards.

## **Data Sources**

Expansion of data linkages and population-based reporting has improved case-finding and enabled surveillance of hemoglobin disorders in both pediatric and adult populations.

Newborn Screening (NBS) cases are linked with live birth records, allowing for linkage with information from death records, Medicaid claims, and immunization records. *These data sources are described below:*

- Birth certificate records have been linked with NBS results for Sickle Cell Disease (SCD) cases for birth years 1988-2011. Since 2008, the linkage match rate has been consistently above 99 percent. The linkage rate for earlier years is 95 percent or higher.
- The Sickle Cell Disease Association of America-Michigan (SCDAA-MI) Chapter has been serving as the contracting agency for diagnostic, medical management, genetic counseling, education, and social work services for patients detected through the NBS process since 1987. The SCDAA-MI provides an annual report to Michigan Department of Community Health (MDCH) each fall with aggregate numbers describing children and adults served through the center. Findings from their 2012 report are included in this document.
- Immunization information from the Michigan Care Improvement Registry (MCIR) was obtained for all cases born from 2004-2008 with a confirmatory diagnosis of SCD via a deterministic linkage with live birth records. The Advisory Committee on Immunization Practices (ACIP) recommended childhood immunization schedule, 2004-2010, was used to determine the appropriate number of vaccines and age of series completion through 18 months.
- Michigan's data warehouse, containing Michigan Medicaid and Children's Special Health Care Services claims, serves as an important source for healthcare and pharmaceutical utilization estimates for both NBS-confirmed individuals with SCD and others not detected through NBS. To further enhance case finding, an algorithm to identify SCD cases using claims was developed. This algorithm is a valuable tool for identifying cases of SCD among those born before 1987 or those not screened in Michigan.

## ***Data Limitations***

- **Newborn screening records:** Only confirmed Sickle Cell Disease (SCD) cases identified through Michigan's Newborn Screening (NBS) Program were included, so any cases not screened in Michigan due to refusal, miss, or migration to Michigan after birth are not captured. Additionally, any infants with an initial screen positive for SCD, but did not have a confirmatory diagnosis reported back to the Program, were excluded.
- **Sickle Cell Disease Association of America (SCDAA):** Data are only generalizable to people who are followed by the SCDAA. The antibiotic use information is based on the date that the prescription was written, so it does not reflect compliance or length of use.
- **Michigan Care Improvement Registry (MCIR):** The quality of the immunization information depends on consistent, accurate reporting by healthcare providers. Not all NBS records could be linked to MCIR records.
- **Michigan Medicaid Claims: The limitations are:**
  - Claims only represent services which were billed and paid for and will not include all services.
  - The data warehouse is populated by claims submitted, so any errors in those claims will be reflected in the data.
  - Children had to be fully-enrolled in a Medicaid program for eleven or more months with no other insurance in a calendar year, so analyses may not be generalized to children enrolled less than eleven months and/or with other insurance
  - The linkage of NBS data to Medicaid claims may have missed people. Although birth certificates were used as the intermediate file when linking NBS and data warehouse records, some NBS records could not be linked to birth certificates and/or data warehouse records due to name changes.
  - Since individuals fulfilling the algorithm criteria of three or more SCD-related claims were included, it is possible that some individuals without SCD met that criteria and were included, but this probability is low based on tests of various algorithms.<sup>4</sup>



## ***Healthy People 2020 Summary of Objectives***

### **Blood Disorders and Blood Safety - Hemoglobinopathies**

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The United States Department of Health and Human Services (HHS) *Healthy People* provides science-based, ten-year national objectives for improving the health of all Americans. For three decades, Healthy People has established benchmarks and monitored progress over time. As a result of stakeholder feedback during the objectives development phase for Healthy People 2020, Blood Disorders and Blood Safety was included in the new initiative. The goal of this objective is to prevent illness and disability related to blood disorders and the use of blood products, including hemoglobinopathies and abnormal bleeding and clotting.<sup>5</sup>

#### **The following *Healthy People* objectives are relevant to Michigan's surveillance baseline measures:**

- Increase the proportion of persons with hemoglobinopathies who receive care in a patient/family-centered medical home.
- Increase the proportion of persons with hemoglobinopathies who receive disease-modifying therapies.
- Increase the proportion of persons with a diagnosis of hemoglobinopathies who receive early and continuous screening for complications.
- Increase the proportion of persons with a diagnosis of hemoglobinopathies and their families who are referred for evaluation and treatment.
- Increase the proportion of children with sickle cell disease who receive penicillin prophylaxis from four months until five years of age.
- Increase the proportion of persons with blood disorders who receive recommended vaccinations.

#### **Additional *Healthy People* objectives are:**

- Increase the proportion of hemoglobinopathy carriers who know their own carrier status.
- Increase the number of community-based organizations (CBOs) that provide outreach and awareness campaigns for hemoglobinopathies.
- Increase the proportion of persons with a diagnosis of hemoglobinopathies who complete high school education or a General Education or Equivalency Diploma (GED) by 25 years of age.



# Incidence

SCD 2014 Surveillance Report

Hemoglobinopathies are a group of inherited disorders in which there is abnormal production or structure of the hemoglobin molecule.<sup>6</sup> Such disorders include hemoglobin C disease, hemoglobin SC disease, Sickle Cell Disease (SCD), and various types of thalassemia. Approximately 300,000 children worldwide are born with this disorder each year.<sup>7</sup>

The primary hemoglobin disorder is SCD, a recessively-inherited disorder mainly found in populations with African, Mediterranean, Middle Eastern, Indian, Asian, Caribbean, and South and Central American ancestry.<sup>8</sup>

## Key Findings:

- From 1988-2011, a total of 1,689 newborns were detected and confirmed with SCD in Michigan.
- In Michigan in 2011:
  - 1 in 364 screened African American newborns and 1 in 1,907 screened newborns (52 per 100,000 screened births) were diagnosed with SCD.
  - An additional 2,817 newborns were identified as having sickle cell trait based on initial screening results.
  - 61 newborns were diagnosed with SCD. Of those, 54 percent were classified as sickle cell anemia (SS), 33 percent as SC disease, and 13 percent as sickle beta thalassemia.

<b>SCD Subtype, Confirmed Cases, 2011</b>		
<b>Subtype</b>	<b>N</b>	<b>%</b>
Hemoglobin SS Disease (HbSS)	33	54
Hemoglobin SC Disease (HbSC)	20	33
Sickle Beta Thalassemia Plus	8	13

Source: Newborn screening follow-up, Michigan Department of Community Health



# Demographics

SCD 2014 Surveillance Report

## Key Findings:

- Nearly all Michigan newborns detected with Sickle Cell Disease (SCD) were black (95 percent of newborns born 1988-2011).
- One in four mothers of Michigan newborns diagnosed with SCD born from 1988-2011 had less than a high school education and only eight percent had at least a college degree at the time of their child's birth.
- Nearly three out of every four Michigan children with SCD resided in the Detroit / Ann Arbor metropolitan area at the time of birth.
- From 1987-2010, newborns diagnosed with SCD lived in five metropolitan areas of the southern and central lower peninsula.

## Demographics of Michigan SCD Cases, Birth Years 1988-2011 (n=1,689)

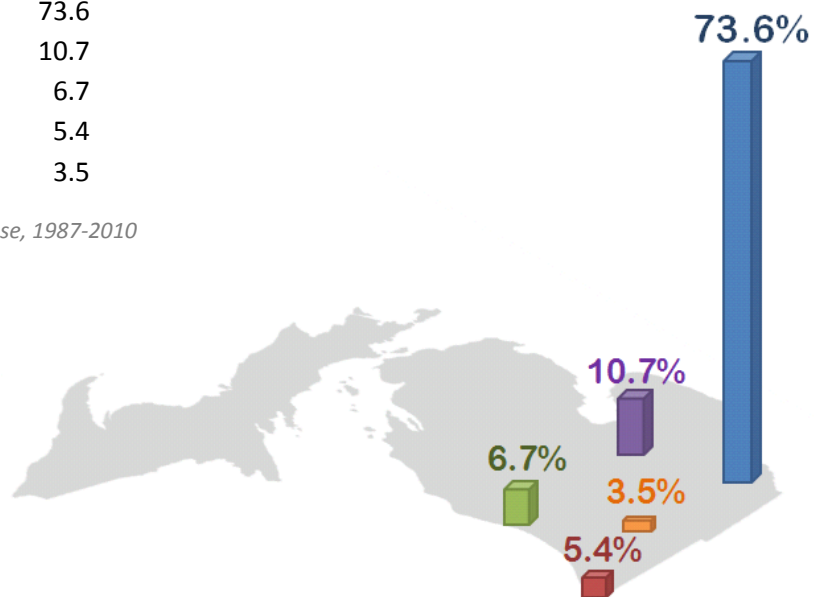
	N	%
<b>Gender</b>		
Female	821	49
Male	868	51
<b>Race</b>		
White	58	3
Black	1605	95
Other	26	2
<b>Education of Mother</b>		
<12th grade, no diploma	429	25
High school diploma or GED	582	34
Some college	367	22
Bachelor's degree	78	5
Graduate degree	47	3
Missing	186	11

Source: Newborn screening follow-up, Michigan Department of Community Health

## Birthplace Region of Michigan SCD Cases, Birth Years 1987-2010

	%
● Detroit & Ann Arbor	73.6
● Saginaw & Flint	10.7
● Grand Rapids & Muskegon	6.7
● Kalamazoo & Benton Harbor	5.4
● Lansing & Jackson	3.5

Source: SCDA-MI newborn screening follow-up database, 1987-2010





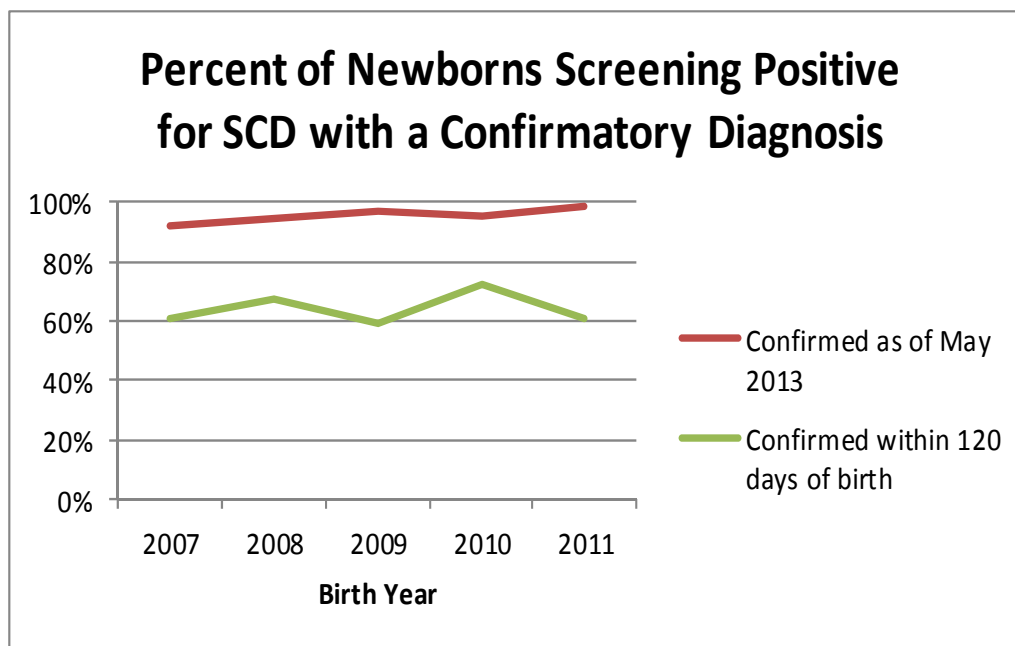
# Confirmatory Diagnosis

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The Michigan Newborn Screening (NBS) laboratory screens for the presence of abnormal hemoglobin in infants using high-performance liquid chromatography and isoelectric focusing. Hemoglobin electrophoresis is recommended for follow-up testing of all positive screens in order to confirm the true diagnosis. The initial screening is performed by the state laboratory, with long-term follow-up for all hemoglobinopathies monitored by the Sickle Cell Disease Association of America, Michigan Chapter through a contractual agreement with the Michigan Department of Community Health. Diagnosed cases confirmed by electrophoresis are reported back to Michigan Department of Community Health.

## Key Findings:

- For birth years 2007-2011, a total of 375 newborns screened positive for Sickle Cell Disease (SCD). A total of 358 received a confirmatory diagnosis (95%); 230 received that diagnosis within 120 days of birth (61%).



Source: Newborn screening follow-up, Michigan Department of Community Health



# Antibiotic Prophylaxis

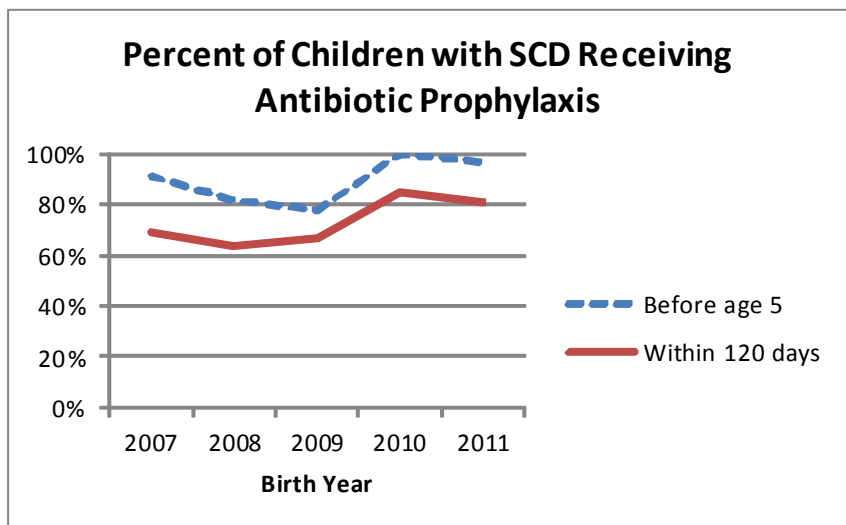
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Before the use of routine penicillin prophylaxis, the case fatality in the United States was as high as 35 percent,<sup>9</sup> with *Streptococcus pneumoniae* infections often progressing quickly to death in less than 24 hours from onset.<sup>10</sup> Historically, the risk of infection among children younger than 5 years of age with Sickle Cell Disease (SCD) has been 3.2 to 6.9 events per 100 patient-years.<sup>11</sup> Following the addition of penicillin prophylaxis for SCD patients younger than five years of age, the rate of infection for *Streptococcus pneumoniae* has decreased to 1.5 events per 100 patient-years.<sup>10</sup> The mortality rate is now between 11 percent and 24 percent and is associated with *Streptococcus pneumoniae* septicemia and meningitis.<sup>12</sup>

The American Academy of Pediatrics recommends the use of penicillin prophylaxis in children with SCD from four months up to age five, and continued prophylaxis in older individuals with a history of severe pneumococcal infection or functional/surgical asplenia. Penicillin prophylaxis for all Michigan children with SCD is monitored by the Sickle Cell Disease Association of America-Michigan Chapter with support from Michigan Department of Community Health.

## Key Findings

- Of the children with SCD born from 2007-2011, 73 percent had treatment initiated within 120 days of birth, and 90 percent had antibiotic prophylaxis at some point before age five years.
- 91 percent of families with children with SCD age four months to five years reported regular administration of penicillin in 2012.



Source: Newborn screening follow-up, Michigan Department of Community Health

## Proportion of Children with SCD age 4 Months to 5 Years Reporting Administration of Penicillin in 2012

Regular administration	216	91%
Non-compliance	3	1%
Unmonitored	18	8%
Total	237	100%

Source: SCDA Annual Report, 9/30/12



# Patient Education

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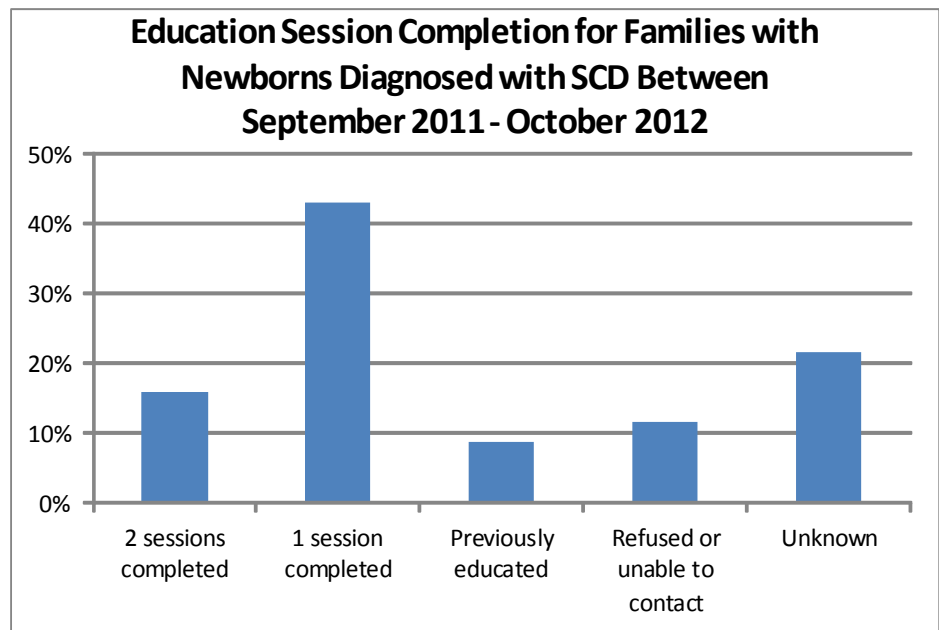
Parental and patient education can significantly reduce morbidity and mortality associated with Sickle Cell Disease (SCD). The Sickle Cell Disease Association of America-Michigan Chapter (SCDAA-MI) educates parents of children with SCD about the diagnosis, symptoms, pain management, and treatments associated with SCD along with best practices for maintaining a healthy lifestyle.

Structured educational sessions are offered for every family of a newborn with SCD. Patient advocates also provide ongoing education for families over the phone, during home visits, and in SCDAA-MI offices located throughout the state. The initial educational process for families of a newborn diagnosed with SCD includes two sessions covering the following topics:

Session 1	Session 2
<ul style="list-style-type: none"> <li>• SCD overview</li> <li>• Health problems that occur early</li> <li>• The difference between sickle cell trait and disease</li> </ul>	<ul style="list-style-type: none"> <li>• Transmission of SCD</li> <li>• Types of SCD</li> <li>• Health problems that occur later in life</li> <li>• SCD and race</li> <li>• Recent strides in SCD research</li> </ul>

## Key findings:

- 16 percent of families completed both educational sessions.
- Approximately two-thirds of the families (67%) received some sort of patient education from the Sickle Cell Disease Association of America.



Source: SCDAA Annual Report, 9/30/12. Data updated 10/15/12.



# Immunizations

SCD 2014 Surveillance Report

Individuals with Sickle Cell Disease (SCD), particularly infants and children, are at higher risk for harmful infections. Pneumonia is one of the leading causes of death in infants and young children with SCD. Comprehensive immunization coverage can reduce the number and burden of infections and their sequelae among children with SCD.

According to the Center for Disease Control & Prevention (CDC), babies and children with SCD should have all of the regular childhood vaccines, plus the following vaccines<sup>13</sup>:

- Influenza every year after six months of age.
- 23-valent pneumococcal vaccine at two and five years of age.
- Meningococcal vaccine (if physician-recommended).

To investigate immunization coverage, records for Michigan children with SCD born 2004-2008 were analyzed in a matched case-control study. Individuals with a confirmatory diagnosis of SCD were compared to disease-free children matched 3:1 on birth month, birth year, and race.

- In 2011, immunization records were retrieved for 262 children with NBS-confirmed SCD and 758 disease-free children.
- Vaccine information came from the Michigan Care Improvement Registry (MCIR).
- The Advisory Committee on Immunization Practices recommended childhood immunization schedule (2004-2010) was used to determine the appropriate number of vaccines and age of series completion through 18 months.

## Key Findings:

- Completion rates for the vaccine series examined varied from 52 percent to 92 percent among SCD cases and 48 percent to 92 percent among non-SCD controls.
- The pneumococcal vaccine series had the lowest completion rate (52 percent of cases and 48 percent of controls) and the hepatitis B series had the highest (92 percent of cases and 91 percent of controls).

Vaccine	SCD Cases*		Controls	
	N	%	N	%
DTaP	166	63.8	413	54.7
Hib	206	79.2	578	76.6
PCV7	135	51.9	360	47.7
Hepatitis B	238	91.5	693	91.8
IPV	223	85.8	626	82.9
MMR	194	74.6	505	66.9
Varicella	185	71.2	499	66.1

\*SCD cases born from 2004-2008 matched to disease-free controls on birth month, birth year, and race

\*\*Defined as age-appropriate series completion by 18 months



# Public Insurance Programs Enrollment

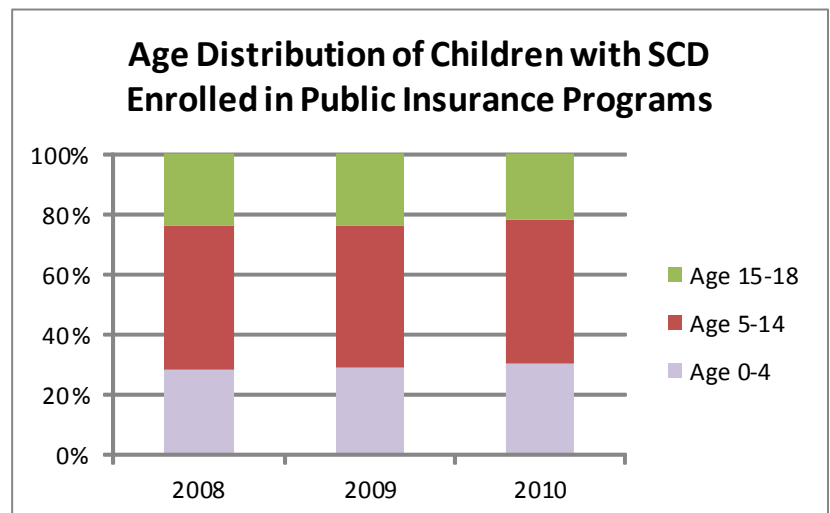
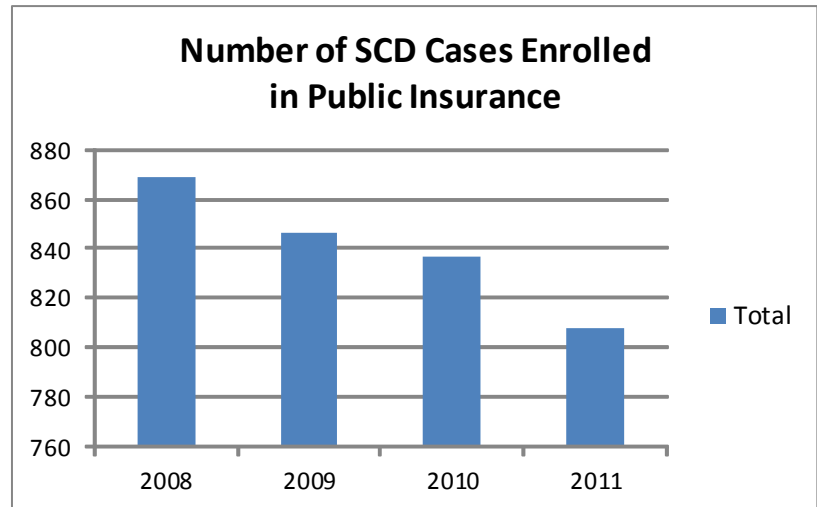
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The Michigan data warehouse contains claims for public insurance programs including Medicaid and Children's Special Health Care Services (CSHCS). All public insurance claims were retrieved for the population of Sickle Cell Disease (SCD) patients up to 18 years of age who were fully enrolled for eleven or twelve months of coverage each calendar year, had no other insurance, and were identified through one of the following methods:

- A confirmed SCD diagnosis through NBS
- At least three SCD-related claims in one year
- Had SCD listed as the CSHCS qualification diagnosis

## Key Findings:

- From 2008-2011, an average of 840 patients with SCD aged 0-18 years were enrolled in Michigan Medicaid programs every year.
- Just under 50 percent of the enrollees were between five and fourteen years of age.







# Transcranial Doppler Screening

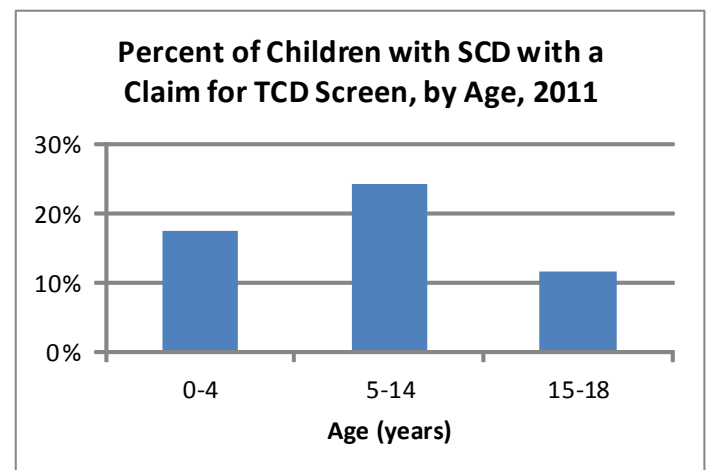
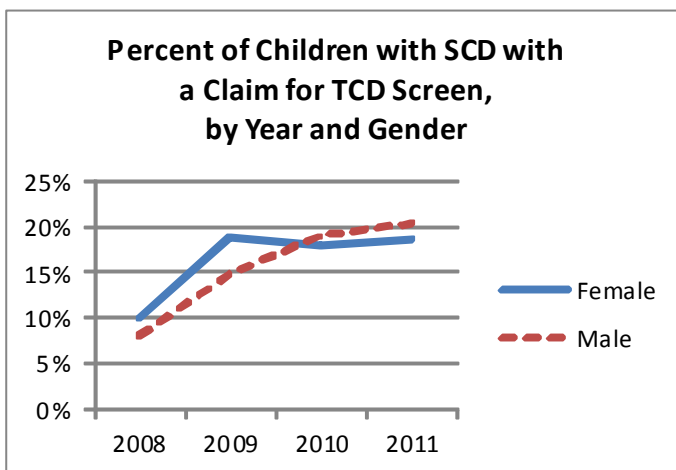
SCD 2014 Surveillance Report

## Transcranial Doppler (TCD) Screening

Sickle Cell Disease (SCD) is associated with a high risk of stroke in the early years of childhood.<sup>14</sup> The risk of a secondary stroke can be significantly reduced through the use of TCD screening. Recommendations for frequency and duration of regular TCD screening vary, but many Michigan physicians agree that TCD screening should be done annually for children with HbSS or HbS-beta thalassemia zero between two and sixteen years of age. Michigan Medicaid claims were examined to determine how many children received TCD screening.

### Key Findings:

- Among children with SCD in Michigan Medicaid programs, the percent with a claim for a TCD screen increased from 2008-2011 among both males and females with approximately 20 percent having a claim for a screen in 2011.
- The oldest age group (15-18 years old) were least likely to have a claim for a TCD screen.





# Hydroxyurea Treatment

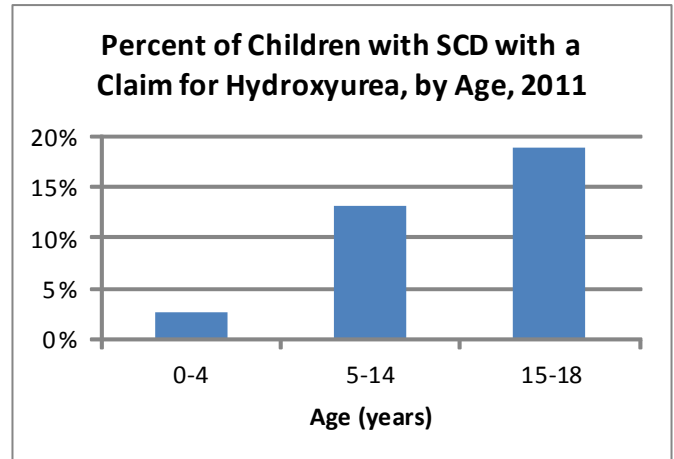
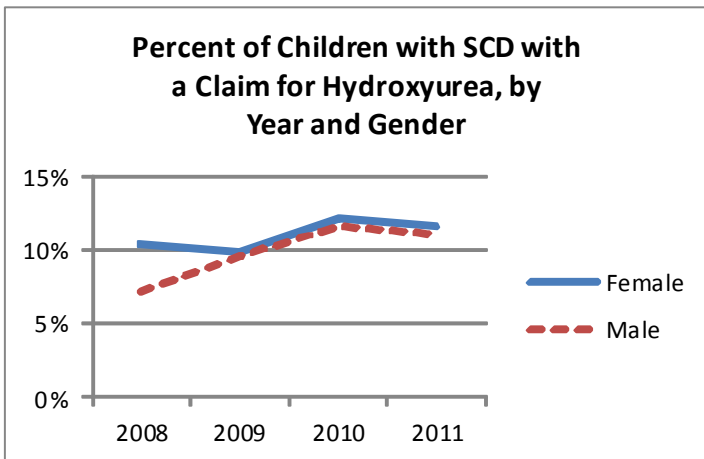
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## Hydroxyurea

Use of hydroxyurea treatment has been shown to reduce the frequency of acute chest syndrome events, pain episodes, blood transfusions, and hospitalizations in individuals with Sickle Cell Disease (SCD).<sup>15</sup> Often, this treatment is considered for patients with severe SCD symptoms. Michigan Medicaid claims were examined to determine how many children with SCD, received hydroxyurea.

### Key Findings:

- Among children with SCD on public insurance, approximately 10-12 percent had a claim for hydroxyurea treatment, with similar coverage for males and females.
- 15-18 year olds are the most common users of hydroxyurea among children with SCD, with 19 percent filling a prescription for hydroxyurea in 2011.





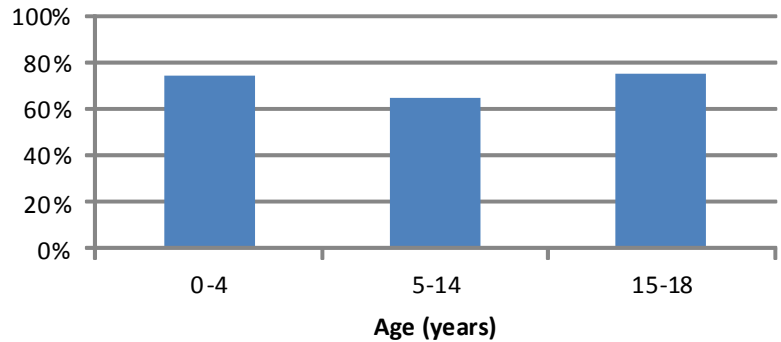
# Emergency Department Visits

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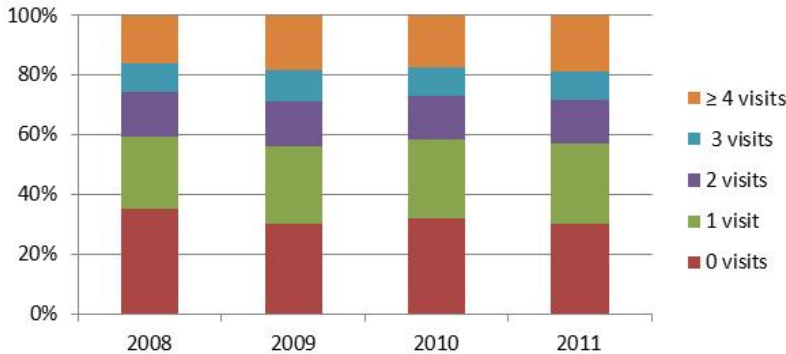
According to the Centers for Disease Control and Prevention (CDC) substantial numbers of emergency department (ED) visits occur among persons with Sickle Cell Disease (SCD). The most common reason for the ED visits for sickle cell patients is pain symptoms.<sup>16</sup>

Repeated ED visits are frequent in the sickle cell population, especially for young adults, who may be particularly at risk as their disease worsens and they transition from pediatric to adult care.

### Percent of Children with SCD with At Least 1 ED Visit During 2011, by Age Category



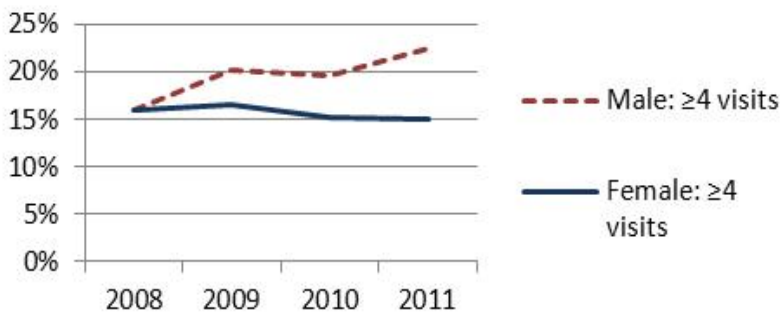
### Number of ED Visits Among Children with SCD



### Key Findings:

- Approximately 75 percent of patients under age five or 15-18 years of age had at least one Medicaid or Children’s Special Health Care Services (CSHCS) claim for an ED visit in 2011, compared to 65 percent of those 5-14 years old.
- Among children with SCD in Medicaid, the percent with at least one ED visit increased from 65 percent in 2008 to 70 percent in 2011. The percent of children with four or more claims for ED visits increased from 16 percent in 2008 to 19 percent in 2011.
- From 2008 to 2011, the proportion of males with SCD with at least four claims for ED visits increased from 16 percent to 22 percent, while the proportion for females remained relatively stable at approximately 15 percent.

### Percent of Children with SCD with 4 or More ED Visits, by Year and Sex



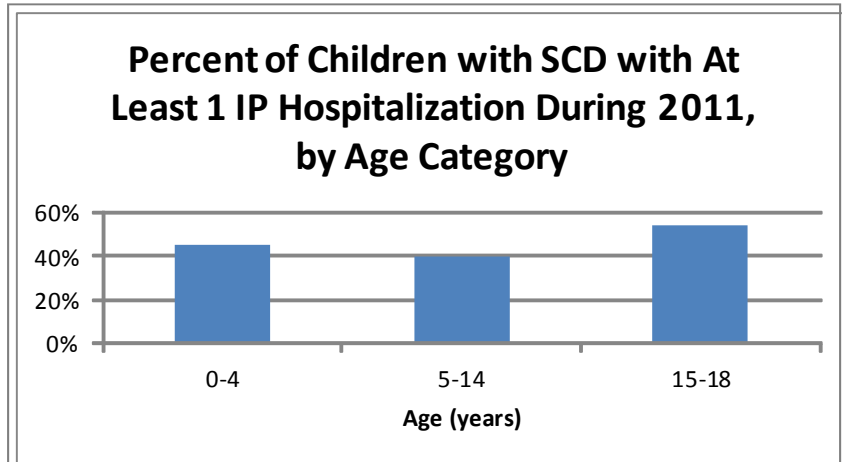


# Inpatient Hospitalizations

SCD 2014 Surveillance Report

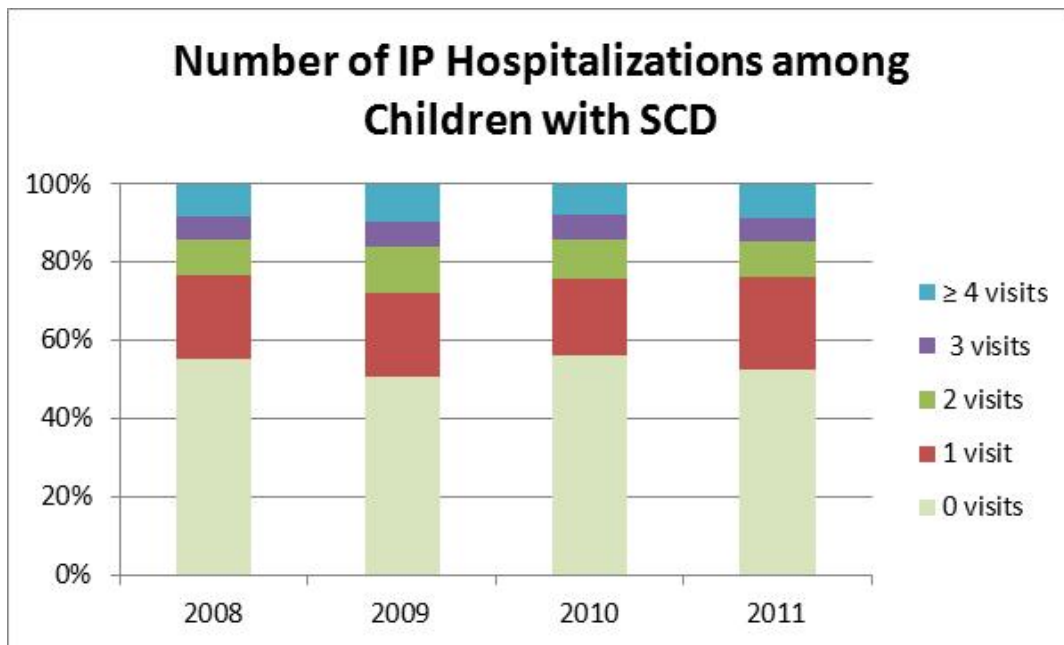
Although Sickle Cell Disease (SCD) is marked by high utilization of medical resources, the full cost of care for patients with SCD is unknown.<sup>17</sup> Sickle cell related complications often require hospitalization.

Interventions designed to prevent SCD complications and avoid inpatient (IP) hospitalizations may also reduce patient rates of re-admission.



## Key Findings:

- Approximately 54 percent of patients 15-18 years of age had at least one claim for an IP hospitalization in 2011, compared to 45 percent of those under five years of age and 40 percent of those 5-14 years old.
- Among children with SCD under 18 years of age enrolled in Medicaid and/or Children’s Special Health Care Services (CSHCS), the percent of children with four or more claims for IP hospitalizations remained relatively stable, ranging from 8-10 percent during the 2008-2011 time period.



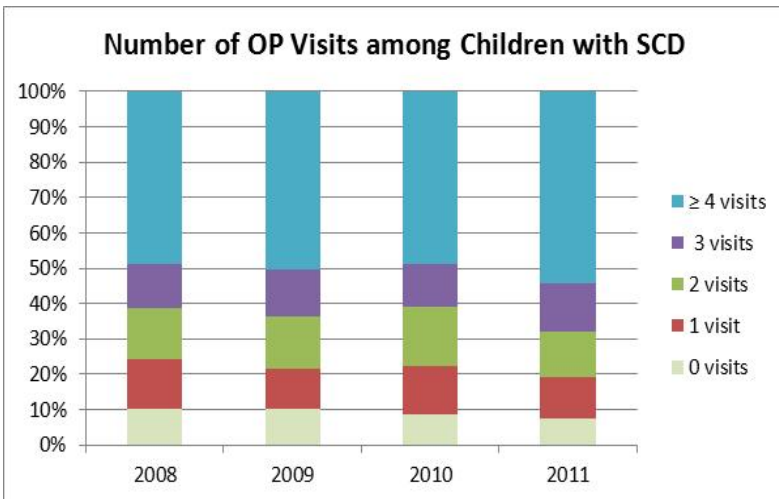
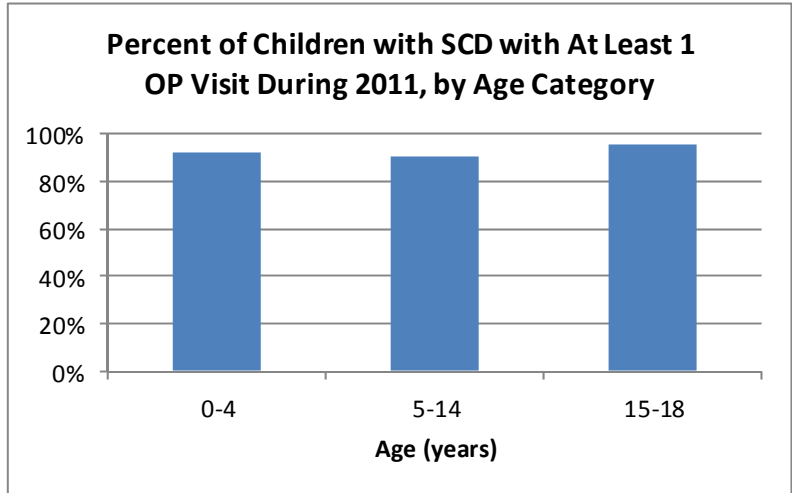


# Outpatient Visits

SCD 2014 Surveillance Report

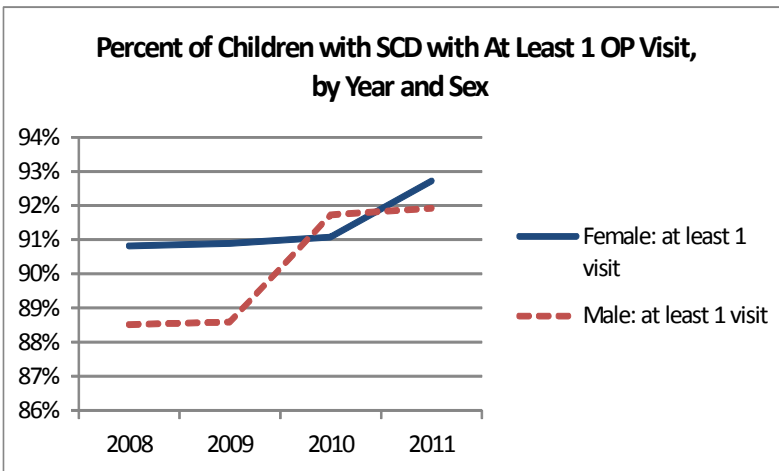
Outpatient (OP) visits may consist of primary care providers and outpatient care by hematologists and other subspecialists. A *Healthy People 2020* objective is to increase the proportion of persons with hemoglobinopathies who receive care in a family-centered medical home.

Outpatient visits may increase likelihood of appropriate referrals, timely immunizations, compliance with penicillin prophylaxis, and reduce need for more costly emergency room care.



### Key Findings:

- Approximately 92 percent of patients under 14 years of age had at least one claim for an OP visit in 2011, compared to 96 percent of those 15-18 years old.
- The percent of children with at least one OP visit increased from 90 percent in 2008 to 92 percent in 2011. The percent of children with four or more claims increased from 49 percent in 2008 to 54 percent in 2011.
- From 2008 to 2011, the proportion of children with Sickle Cell Disease (SCD) with at least one claim for OP visits increased from 89 percent to 92 percent for males and 91 percent to 93 percent for females.



## ***Future Directions***

The Michigan Hemoglobinopathy Quality Improvement Committee (MiHemQIC), in partnership with the Michigan Department of Community Health (MDCH) sickle cell planning team, are building on the work in this report by continuing to monitor short-term follow-up indicators over time, and measuring long-term objectives over time to identify trends in quality of care to create benchmarks for national standards. This work will inform development of a public health plan to address the burden of sickle cell disease (SCD) across the life span. Although life expectancy for individuals with SCD has increased significantly during the past 40 years, the treatment of this disease and resulting reduction of the disease burden has not been adequately addressed.

### **MDCH will engage in a mixture of activities that includes:**

- Enhancing local community networks and partnerships to improve the quality of life for individuals with SCD and their families.
- Implementing prevention strategies to reduce complications in individuals living with SCD.
- Increasing awareness amongst medical and allied health care professionals regarding the needs and experiences of those living with SCD in order to bolster the provision of optimal care to these individuals.
- Collaborating with hospitals and other service providers in order to design and implement projects that will improve access, treatment, and education on the issue of SCD.
- Collaborating with the MiHemQIC and Children's Special Health Care Services (CSHCS) to establish protocols for transitioning patients with SCD from pediatric to adult care and train/support providers in implementing the transitional process.

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