Sickle Cell Disease
Newborn Screening Surveillance






October 2014

Celebrating
27 *Years*
of Michigan Sickle Cell Screening

Background

- Newborn screening (NBS) for sickle cell disease (SCD) has been conducted at the Michigan Department of Community Health since 1987.
- Longitudinal follow-up of patients with SCD diagnosed through NBS is carried out through five years of age by NBS Program staff in collaboration with the Sickle Cell Disease Association of America, Michigan Chapter (SCDAA-MI).
- In 2009, Michigan's NBS Program received a grant* to assess the burden of hemoglobinopathies in Michigan through ongoing surveillance.
- Conducted analysis for the University of Michigan's Child Health Evaluation and Research Unit in support of AHRQ grant

*Funding provided by the Centers for Disease Control and Prevention (CDC) and the National Heart, Lung, and Blood Institute (NHLBI) at the National Institutes of Health (NIH)

Surveillance Objectives

- Monitor short-term patient follow-up indicators over time
- Measure long-term trends in quality of care and outcomes specific to the population with hemoglobinopathies in Michigan
- Create benchmarks for comparison with national standards

Data Sources

- **Newborn Screening Records:**
  - All infants with a positive screen for SCD between birth years 1988-2011

- **SCDAA-MI provides:**
  - Individual-level variables to the NBS Follow-up Program:
    - Confirmatory diagnosis
    - Date of confirmatory diagnosis
    - Date of penicillin prophylaxis initiation
  - A report with aggregate numbers of children and families receiving education

Data Sources

- **Birth Certificate Records:**
  - NBS records for all infants confirmed with SCD between birth years 1988-2011were linked to birth certificate records.
- **Michigan Care Improvement Registry (MCIR):**
  - Immunization information was obtained for SCD cases born 2004-2008
- **Data Warehouse:**
  - Contains Medicaid and Children's Special Health Care Services (CHSCS) records

Data Limitations

- **NBS**
  - Only confirmed SCD cases identified through Michigan's NBS Program were included
- **SCDAA-MI**
  - Penicillin initiation is based on date the prescription was written
- **MCIR**
  - Quality depends on consistent, accurate reporting of immunizations by health care providers

Data Limitations

- **Data Warehouse**
  - Some NBS records could not be linked due to name changes.
  - Since individuals fulfilling the algorithm criteria of 3 or more SCD-related claims were included, some individuals without SCD may have been included.
  - Children had to be fully-enrolled in a Medicaid program for 11 or more months with no other insurance in a calendar year, so analyses may not be generalizable to all children with SCD.
  - 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.

Incidence

- From 1988-2011, 1,689 newborns were detected and confirmed with SCD in Michigan.

- In 2011:
  - 1 in 364 screened African American newborns and 1 in 1,907 screened newborns 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.

SCD Subtype, Confirmed Cases, 2011		
Subtype	**N**	**%**
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

Sickle Cell Disease in Michigan

Demographics of SCD Cases born 1988-2011 (n = 1,689)

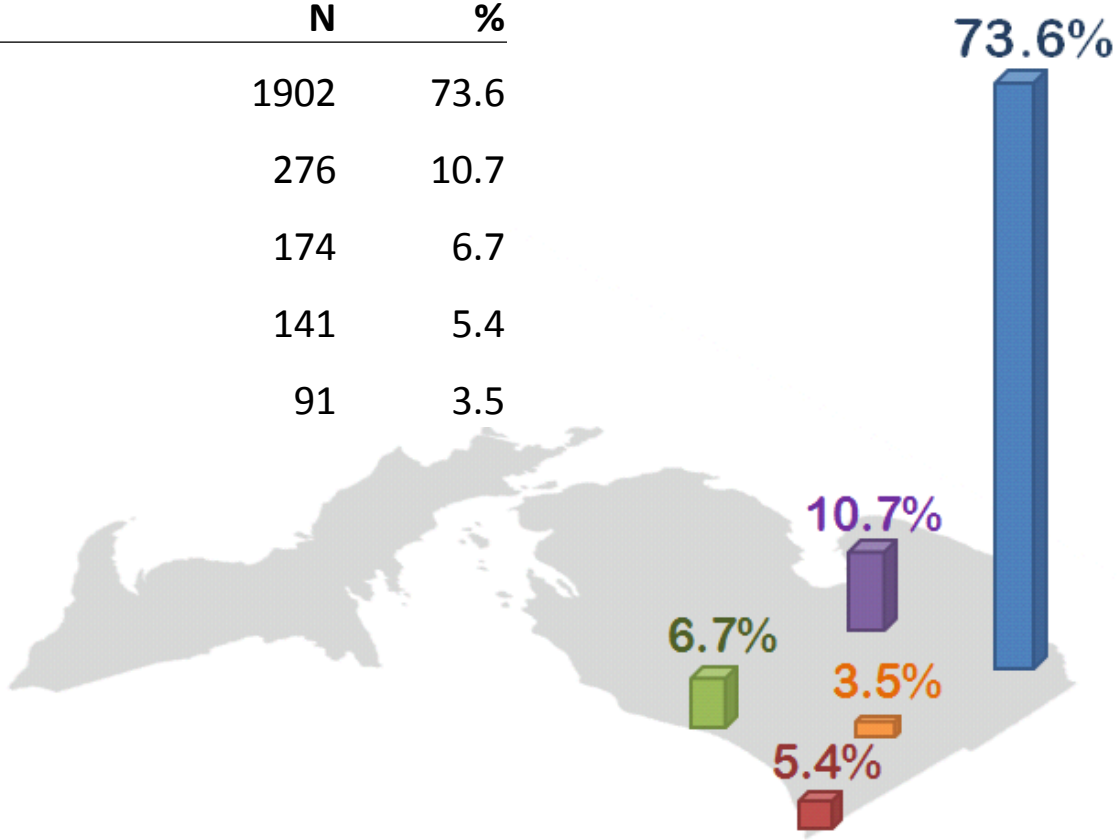
	N	%
Gender		
Female	821	49
Male	868	51
Race		
White	58	3
Black	1605	95
Other	26	2
Education of Mother		
<12th grade, no diploma	429	25
High school diploma or GED equivalent	582	34
Some college, no diploma or Associate's degree	367	22
Bachelor's degree	78	5
Graduate degree	47	3
Missing	186	11

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

Sickle Cell Disease in Michigan

Birthplace of SCD Cases born 1987-2010

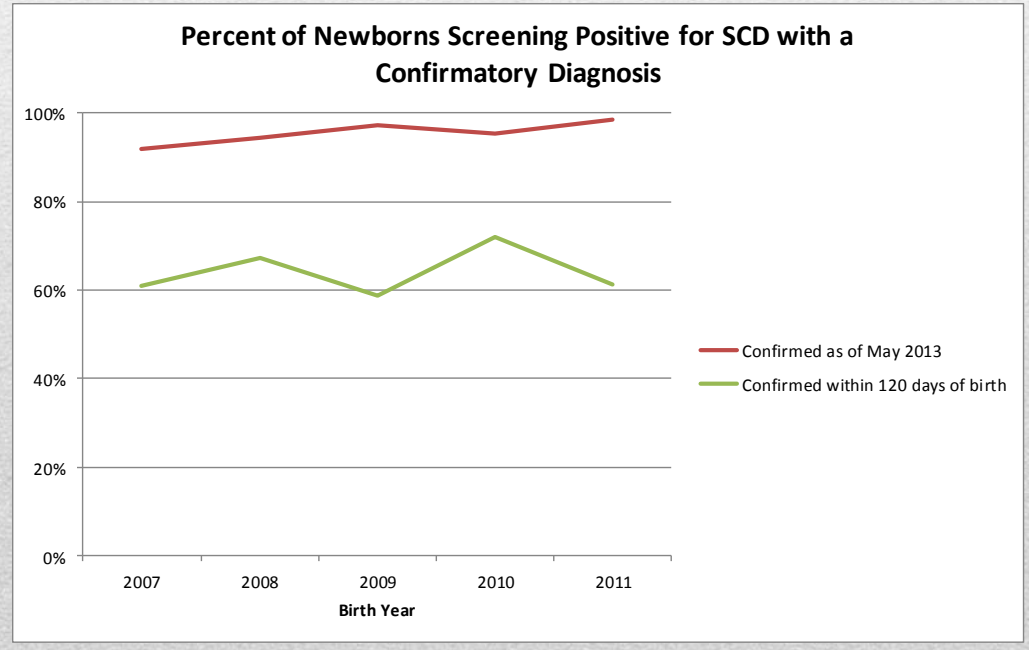
	N	%
Detroit & Ann Arbor	1902	73.6
Saginaw & Flint	276	10.7
Grand Rapids & Muskegon	174	6.7
Kalamazoo & Benton Harbor	141	5.4
Lansing & Jackson	91	3.5



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

Confirmatory Diagnosis

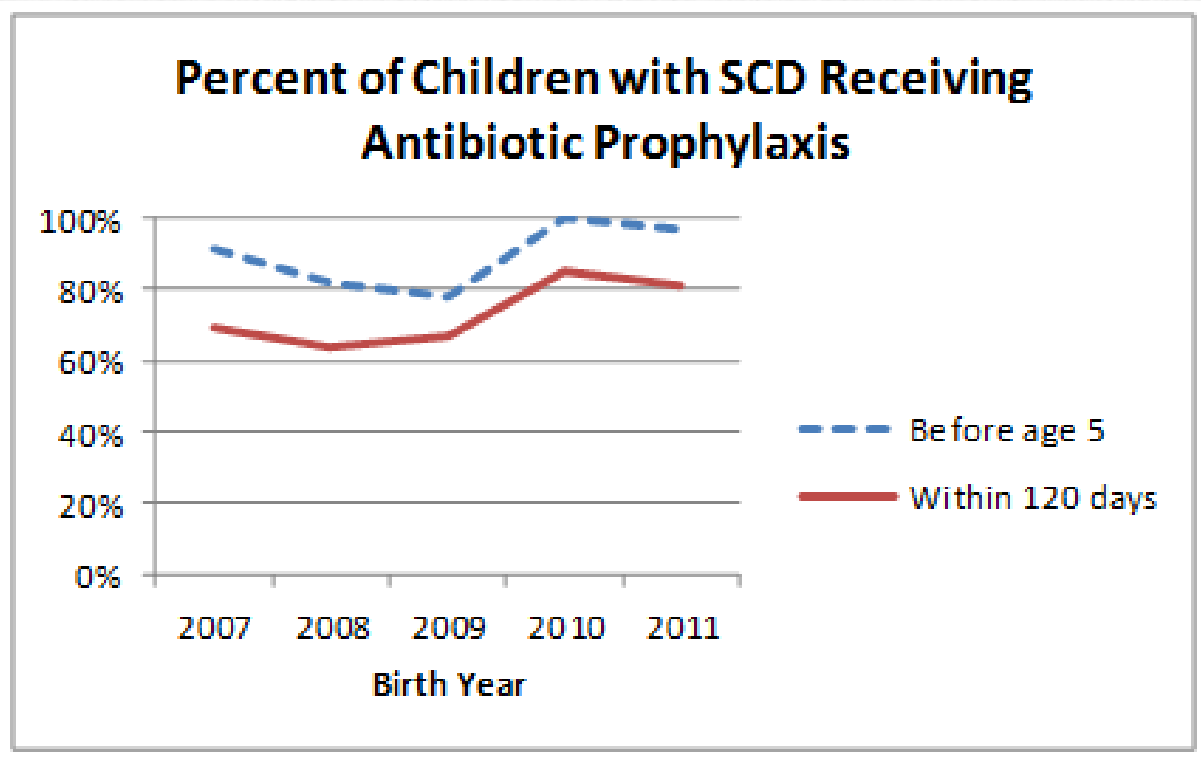
- The Michigan NBS laboratory screens for the presence of abnormal hemoglobin in newborns using high-performance liquid chromatography and isoelectric focusing.
- Hemoglobin electrophoresis is recommended for confirmatory testing of all positive screens in order to verify the true diagnosis.



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

Antibiotic Prophylaxis

- The American Academy of Pediatrics recommends the use of penicillin prophylaxis in children with SCD from four month up to age five.

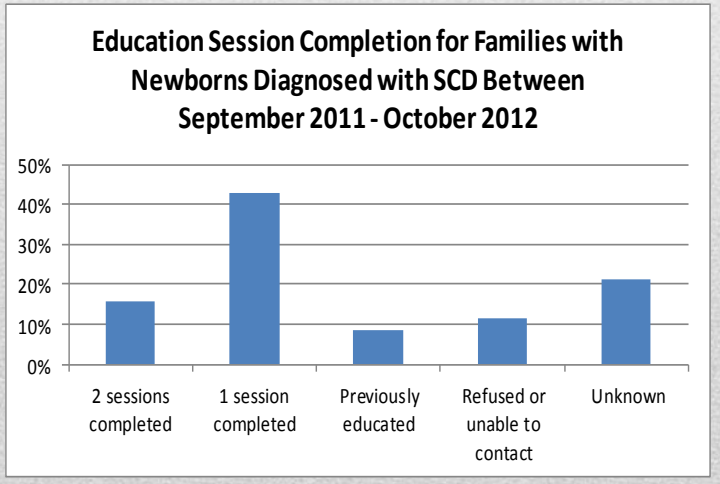


Source: Newborn screening follow-up, Michigan Department of Community Health

Patient Education

- SCDAA's initial educational process for families of a newborn diagnosed with SCD includes two sessions:

Session 1	Session 2
• SCD overview • Health problems that occur early • The difference between sickle cell trait and disease	• Transmission of SCD • Types of SCD • Health problems that occur later in life • SCD and race • Recent strides in SCD research



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

Immunizations

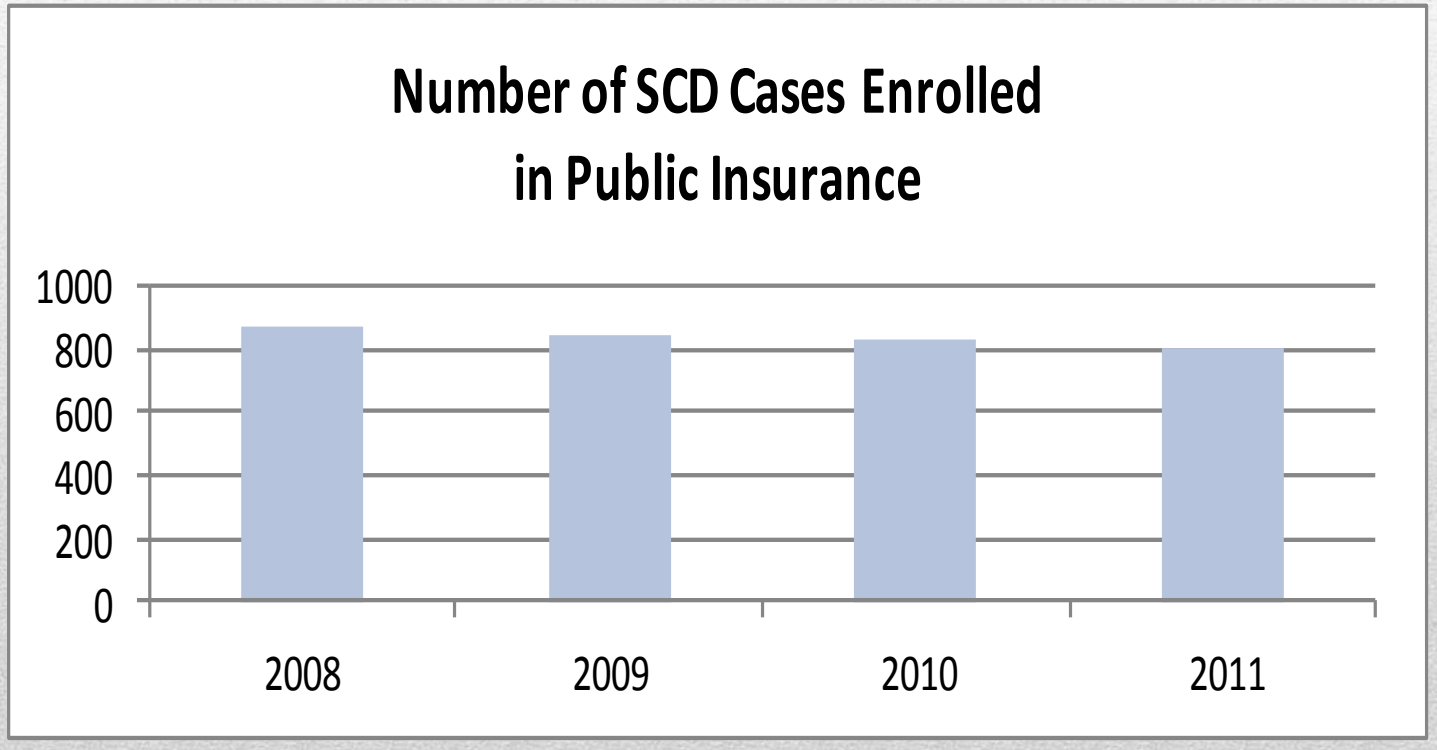
- In 2011, immunization records from the MCIR were retrieved for 262 children with NBS-confirmed SCD and 758 disease-free children matched on birth month/year and race.
- The Advisory Committee on Immunization Practices recommended childhood immunization schedule (2004-2010), was used to determine the appropriate number of vaccines and age of series completion through 18 months.

	SCD Cases		Controls	
	Completed Series		Completed Series	
Vaccine	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

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

Medicaid Programs Enrollment

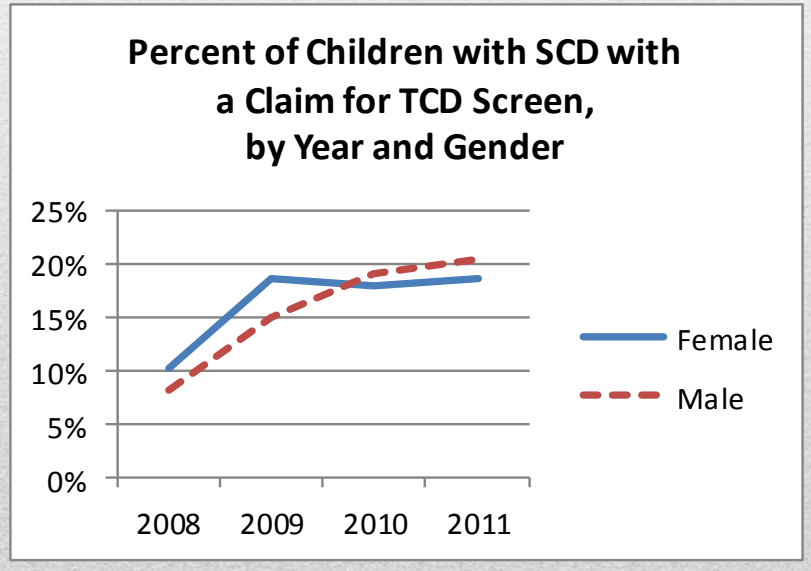
- From 2008-2011, an average of 840 patients with SCD aged 0-18 years were enrolled in Michigan Medicaid programs every year.

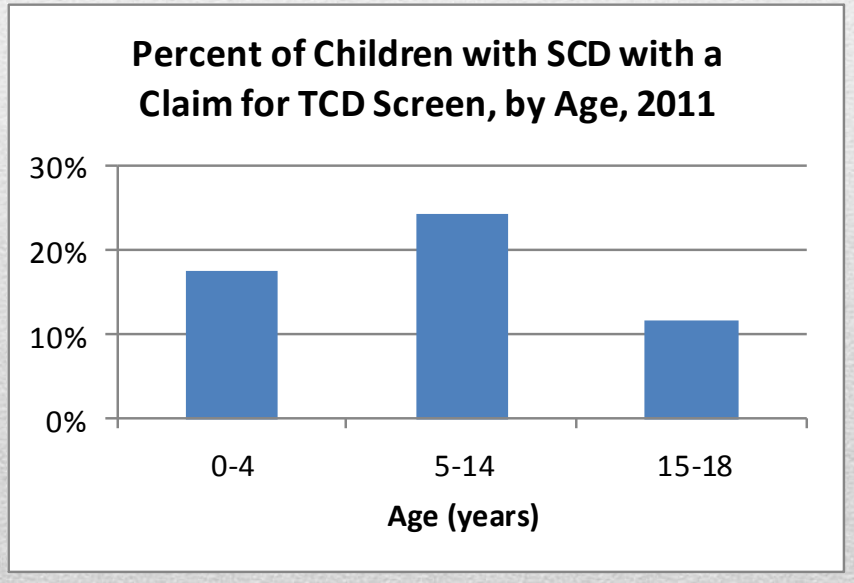


Source: Data warehouse, Michigan Department of Community Health

Transcranial Doppler (TCD) Screening

- The risk of a secondary stroke for children with SCD can be significantly reduced through the use of TCD screening.
- Michigan data warehouse claims were examined to determine how many children with SCD received TCD screening.

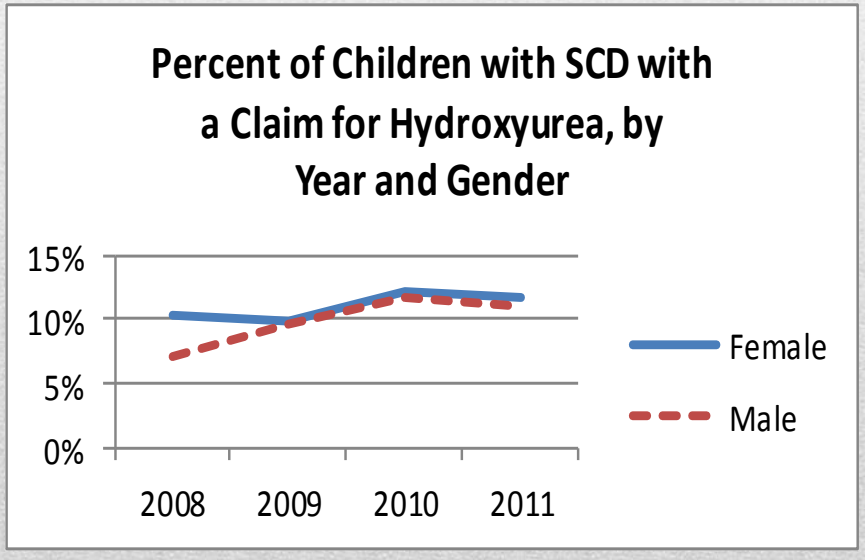


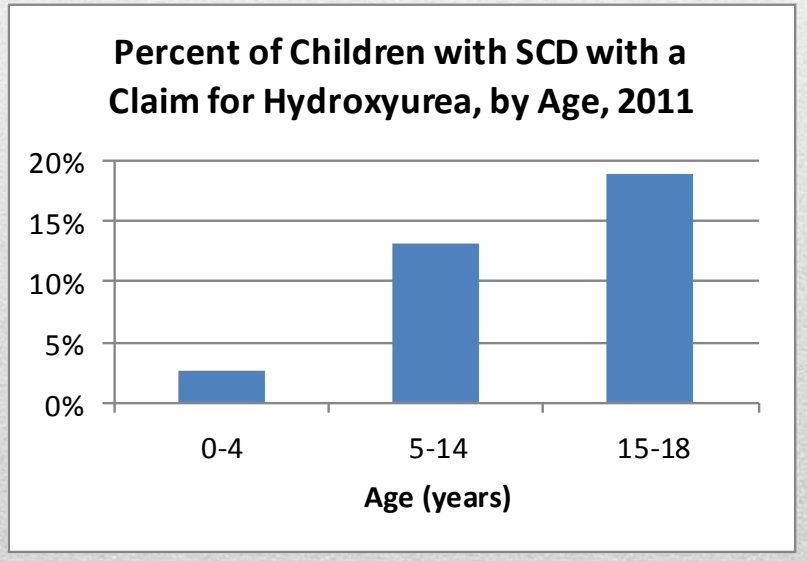


Source: Data warehouse, Michigan Department of Community Health

Hydroxyurea Treatment

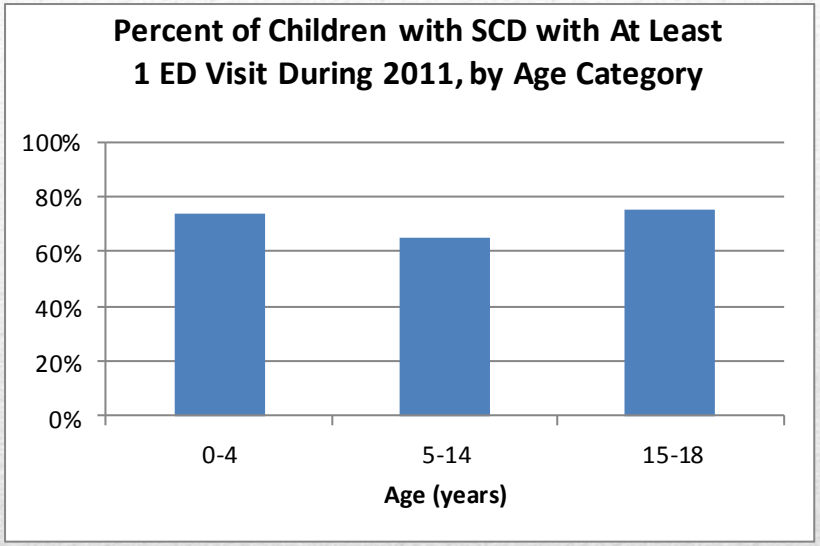
- Use of hydroxyurea treatment has been shown to reduce the frequency of acute chest syndrome events, pain episodes, blood transfusions, and hospitalizations in individuals with SCD.
- Michigan data warehouse claims were examined to determine how many children with SCD received hydroxyurea.

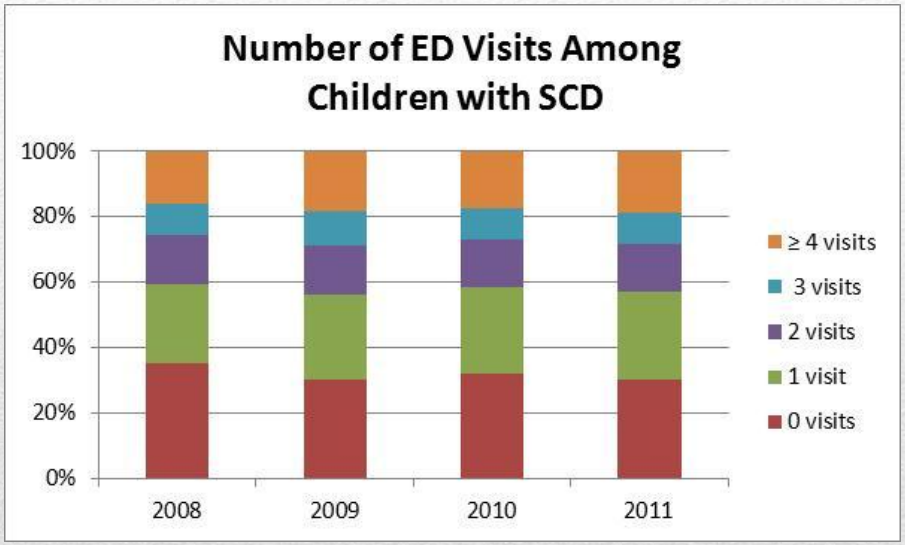




Source: Data warehouse, Michigan Department of Community Health

Emergency Department (ED) Visits





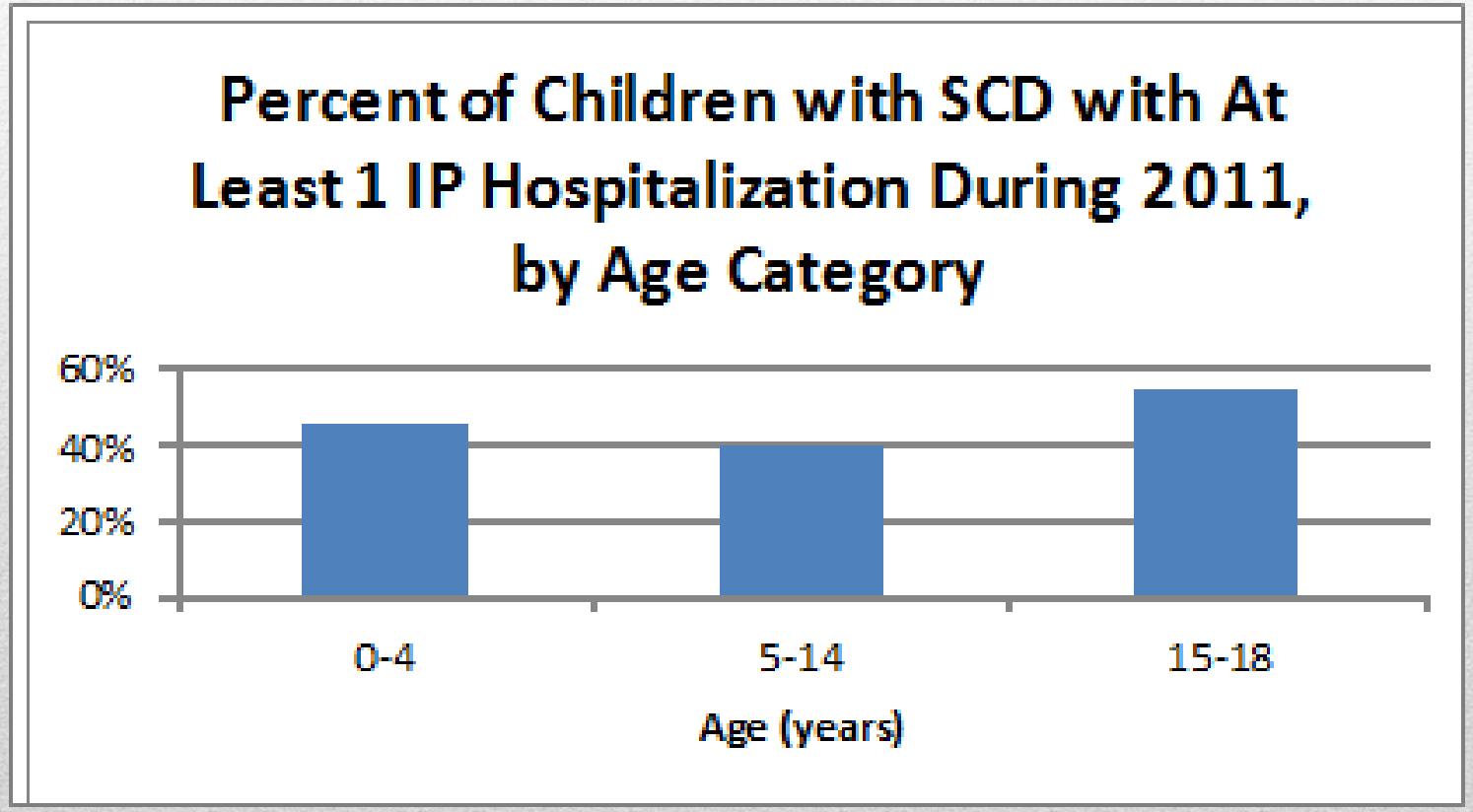
- Approximately 75% of patients under age 5 or 15-18 years of age had at least one Medicaid or CSHCS claim for an ED visit in 2011, compared to 65% of those 5-14 years old.

- Among children with SCD in Medicaid, the percent with at least one ED visit increased from 65% in 2008 to 70% in 2011. The percent of children with 4 or more claims for ED visits increased from 16% in 2008 to 19% in 2011.

Source: Data warehouse, Michigan Department of Community Health

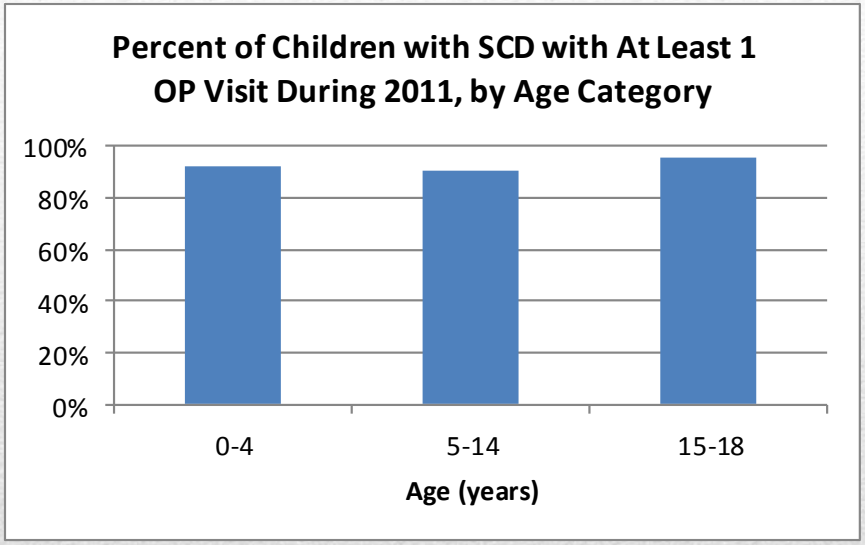
Inpatient (IP) Hospitalizations

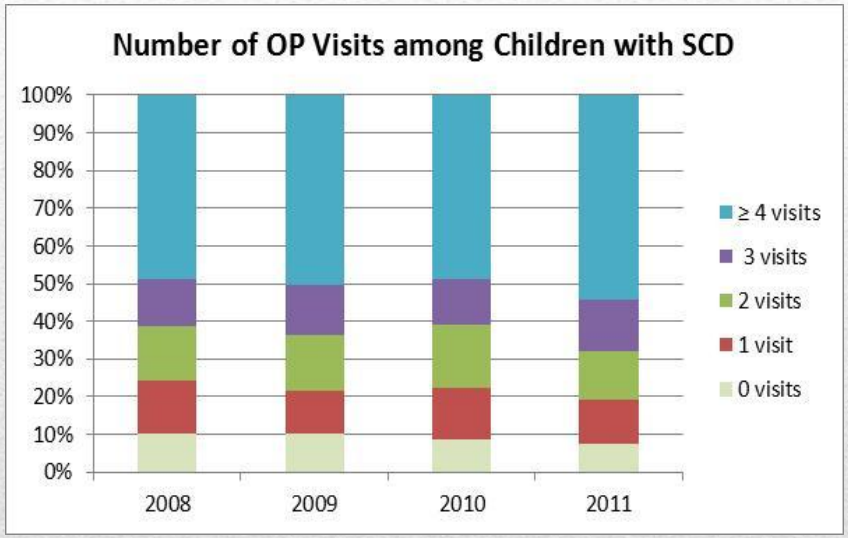
- Approximately 54% of patients 15-18 years of age had at least one claim for an IP hospitalization in 2011, compared to 45% of those under 5 years of age and 40% of those 5-14 years old.



Source: Data warehouse, Michigan Department of Community Health

Outpatient (OP) Visits





- Approximately 92% of patients under 14 years of age had at least one claim for an OP visit in 2011, compared to 96% of those 15-18 years old.

- The percent of children with at least one OP visit increased from 90% in 2008 to 92% in 2011. The percent of children with 4 or more claims increased from 49% in 2008 to 54% in 2011.

Source: Data warehouse, Michigan Department of Community Health

Future Directions

- Continue surveillance of incidence, short term outcomes, health care utilization, and impact of disease on children's lives.
- Enhance community networks and partnerships to improve the quality of life for individuals with SCD and their families.
- Increase 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.
- Collaborate 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.