



SICKLE CELL DISEASE SURVEILLANCE IN CALIFORNIA: METHODS, FINDINGS, AND CHALLENGES

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Background

Registry & Surveillance for Hemoglobinopathies (RuSH)

- Funded by NIH-National Heart, Lung & Blood Institute
- 7 State Collaborative (CA, GA, FL, MI, NC, NY, and PA)
- Project Goal:
 - Develop a strategy to count people with sickle cell disease
 - Describe the population of affected individuals
 - Connect to community stakeholders

California RuSH Project

Created a profile/snapshot of the population with SCD (2004-2008 data)

- Incidence among new births
- Prevalence in general population
- Demographic characteristics of the population: age, sex, race
- Geographic distribution
- Mortality rates
- Health care utilization: hospitalizations, emergency department visits, disease complications & procedures
- Describe services provided to newborns at specialty care centers in the first five years of life

California RuSH Project

Collected data from different sources:

- Newborn Screening Program (2000-2008)
- Medi-Cal (Medicaid) (2000-2008)
- Hospital Discharge (2004-2008)
- Emergency Room Admissions (2005-2008)
- Vital Records – Birth (2000-2008)
- Vital Records – Death (2004-2008)
- CHLA (incl. adults) (2000-2008)
- CHRCO (incl. adults) (2000-2008)

RuSH Project

CDC's RuSH Surveillance Design Team developed a case definition to determine the level of certainty of diagnoses.

- *Level 1: ‘**Confirmed**’ cases came from NBS and Clinic Data where cases had a specific genotype of SCD.*
- *Level 2: ‘**Probable**’ cases had two or more encounters with a relevant ICD 9 diagnostic code across administrative data sources in combination with one or more designated complications, procedures or treatments.*
- *Level 3: ‘**Possible**’ cases were those not meeting above criteria but seen with relevant ICD 9 codes in any data source.*

Complications, Procedures, and Treatments

Complications

- ❑ Chronic Renal failure
- ❑ Pneumonia, ACS
- ❑ Pulmonary hypertension
- ❑ Stroke
- ❑ Intracranial bleeding
- ❑ Priapism
- ❑ Iron overload
- ❑ Gallstones/cholelithiasis, cholecystitis
- ❑ Avascular necrosis
- ❑ Retinal disease
- ❑ Splenomegaly, splenic sequestration, hypersplenism
- ❑ Leg ulcers
- ❑ Dactylitis
- ❑ Osteomyelitis

Procedures

- ❑ Red Cell Transfusion
- ❑ Red Cell Exchange
- ❑ Splenectomy
- ❑ Cholecystectomy
- ❑ Transcranial Doppler

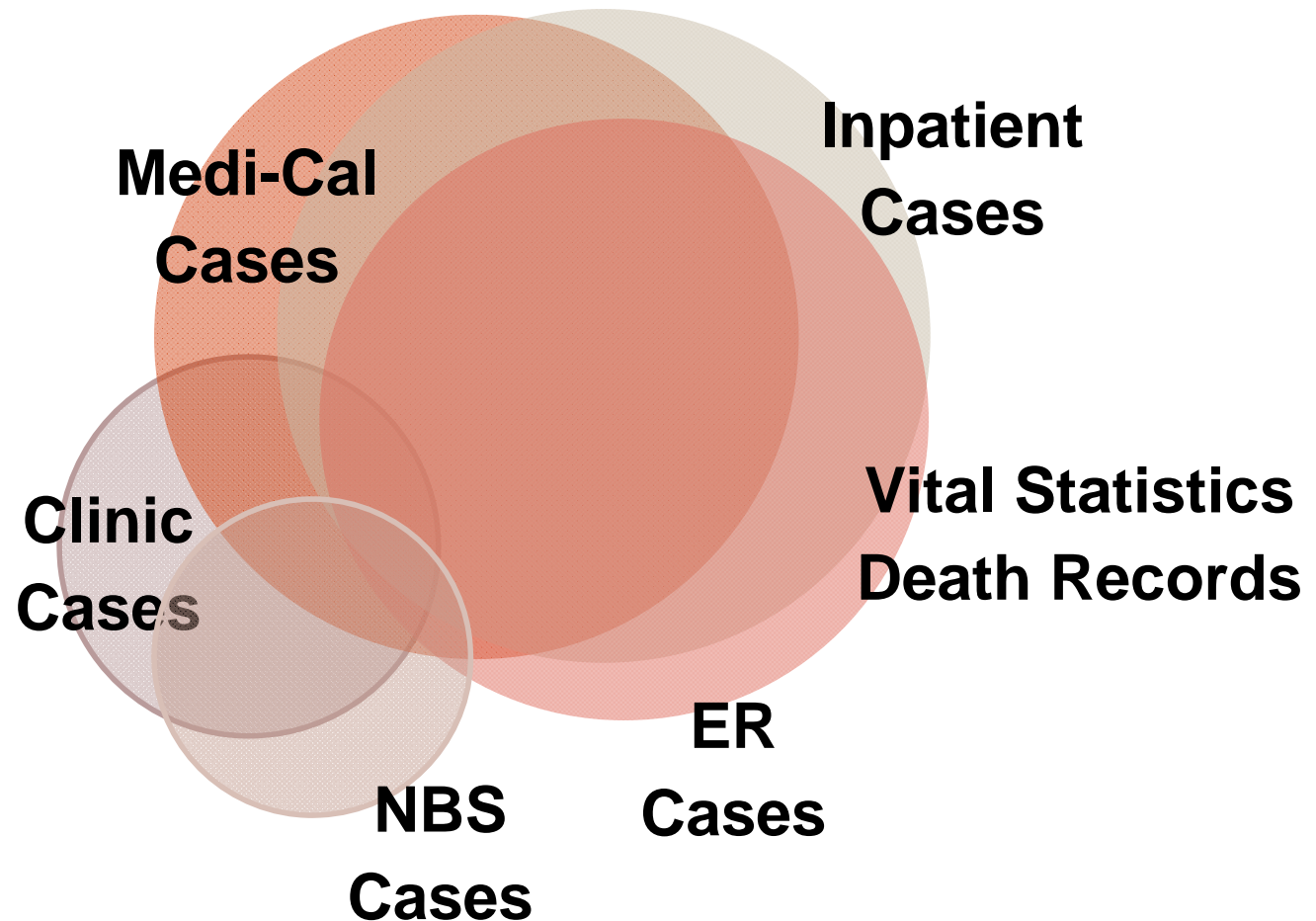
Treatments

- ❑ Hydroxyurea
- ❑ Parenteral Analgesics
- ❑ Iron Chelators
- ❑ Erythropoietin
- ❑ Folic Acid

Linkage Methods

- Goal: Capture each individual just one time
- Exact matching where SSN/sex/diagnosis match.
- Where no SSN or above do not match, other variables included:
 - Diagnosis (based on confirmed NBS or clinic diagnosis or ICD 9 code from administrative data);
 - Date of birth (with weighting for close match);
 - Sex;
 - First 3 digits of zip code; and
 - Hospital facility, when available.

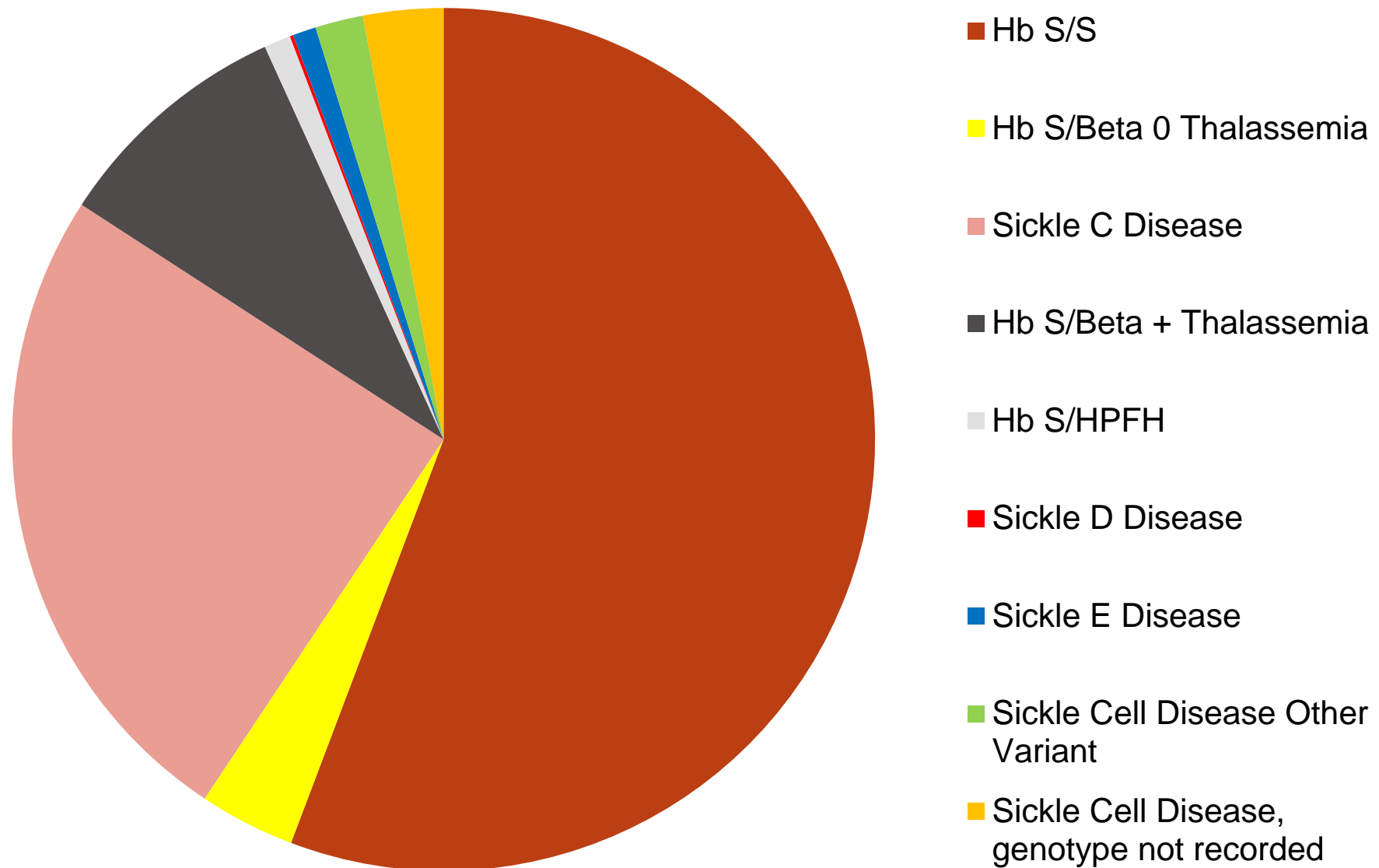
Data Sources For Case Finding Overlapped



RuSH Project Findings

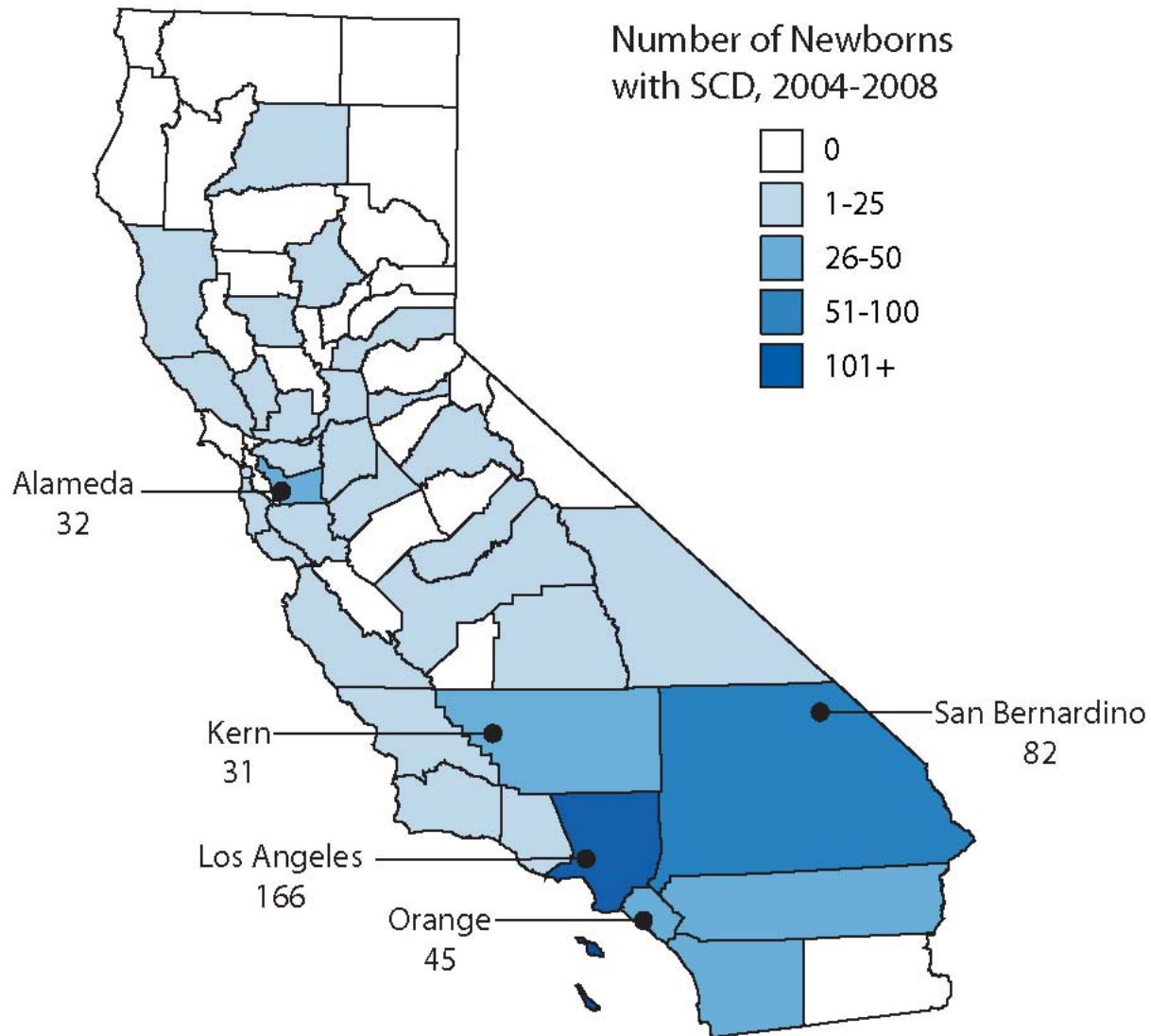
- We identified 6,207 people with sickle cell disease in California between 2004-2008
- 2,397 confirmed and 3,810 probable SCD cases
 - 43% were younger than 18 years of age
 - 21% were 18 to 29 years
 - 25% were 30 to 50 years
 - 11% were 51 years or older
- 1 out of every 8,000 live births in the California general population
- 1 out of 500 live births of Black or African Americans
- 1 out of 99,000 live births of Hispanic Americans.

Distribution of Confirmed SCD Cases (n= 2,397)

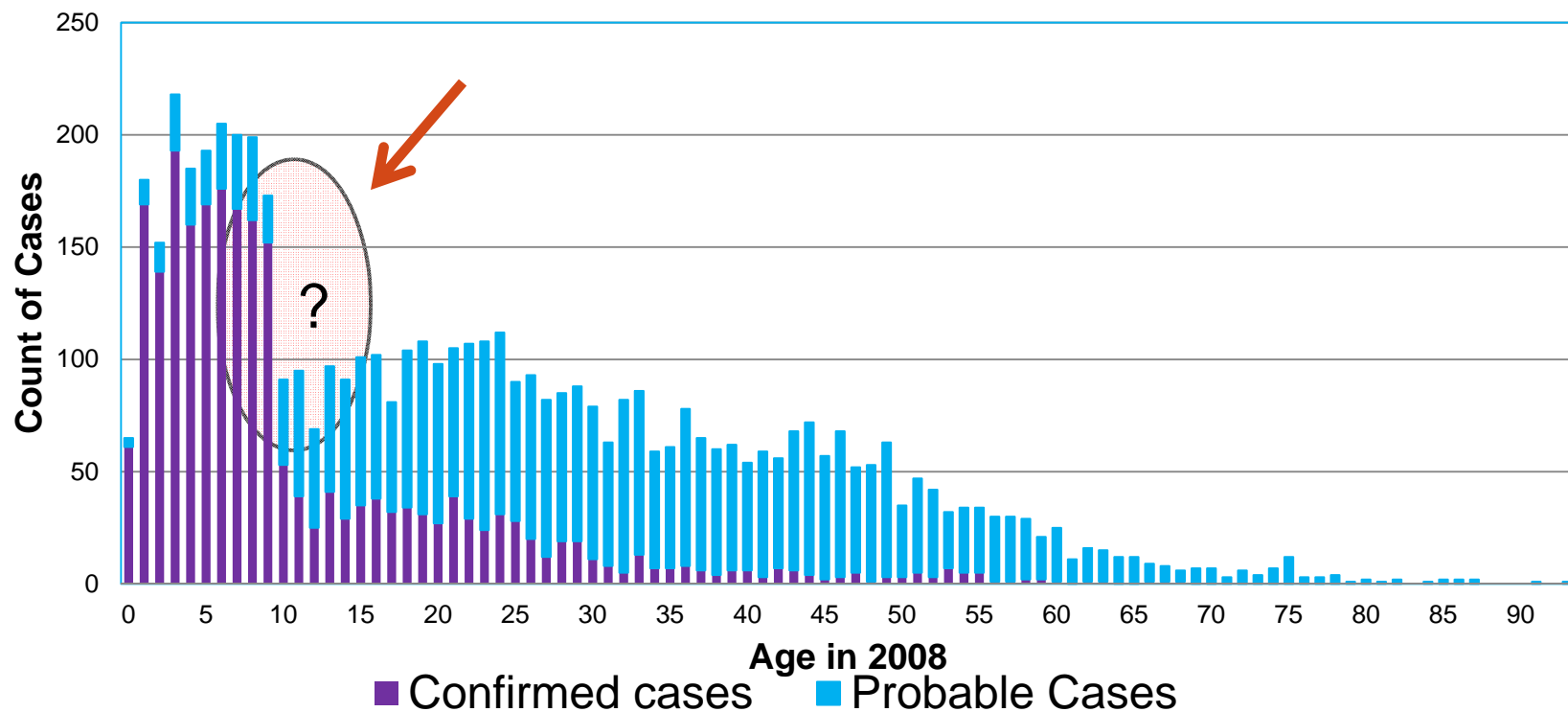


SCD Cases Among Live Births by County: 2004-2008

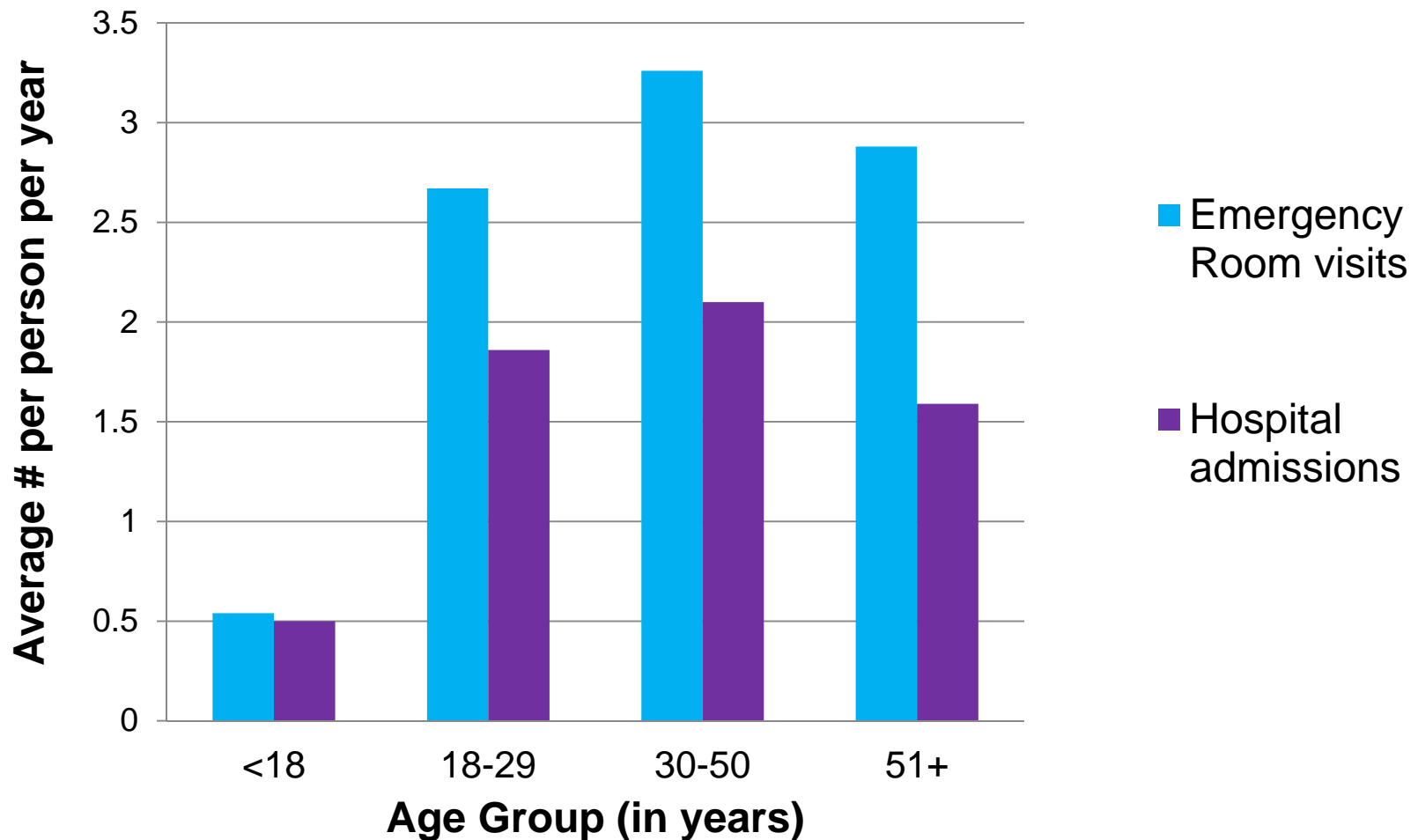
n=1,472



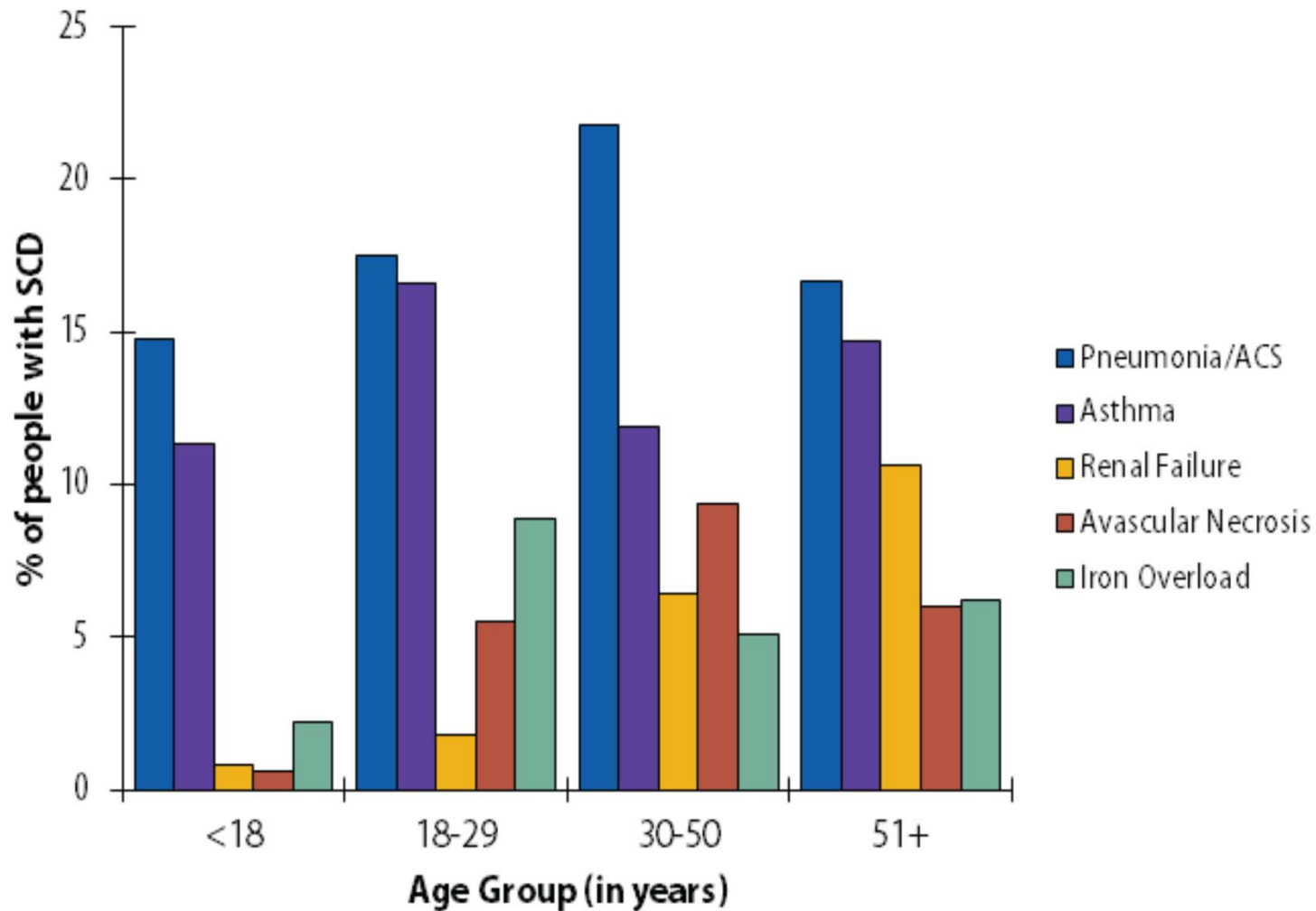
Age Distribution of Confirmed/Probable SCD Cases (average age = 22.8 years)



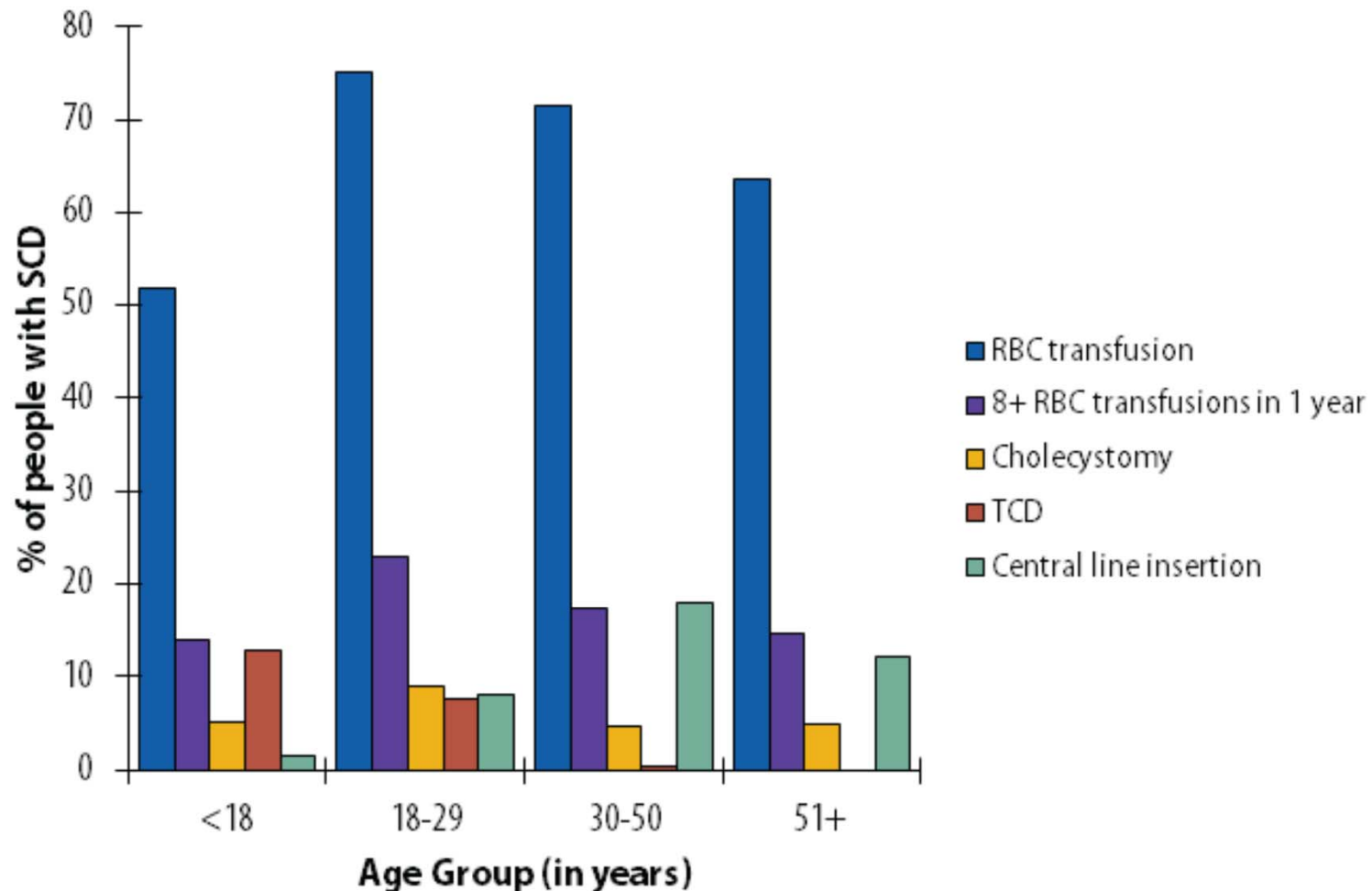
Emergency Department Visits & Inpatient Hospitalizations: 2004-2008



Five most common complications for person with SCD on Medi-Cal: 2004-2008

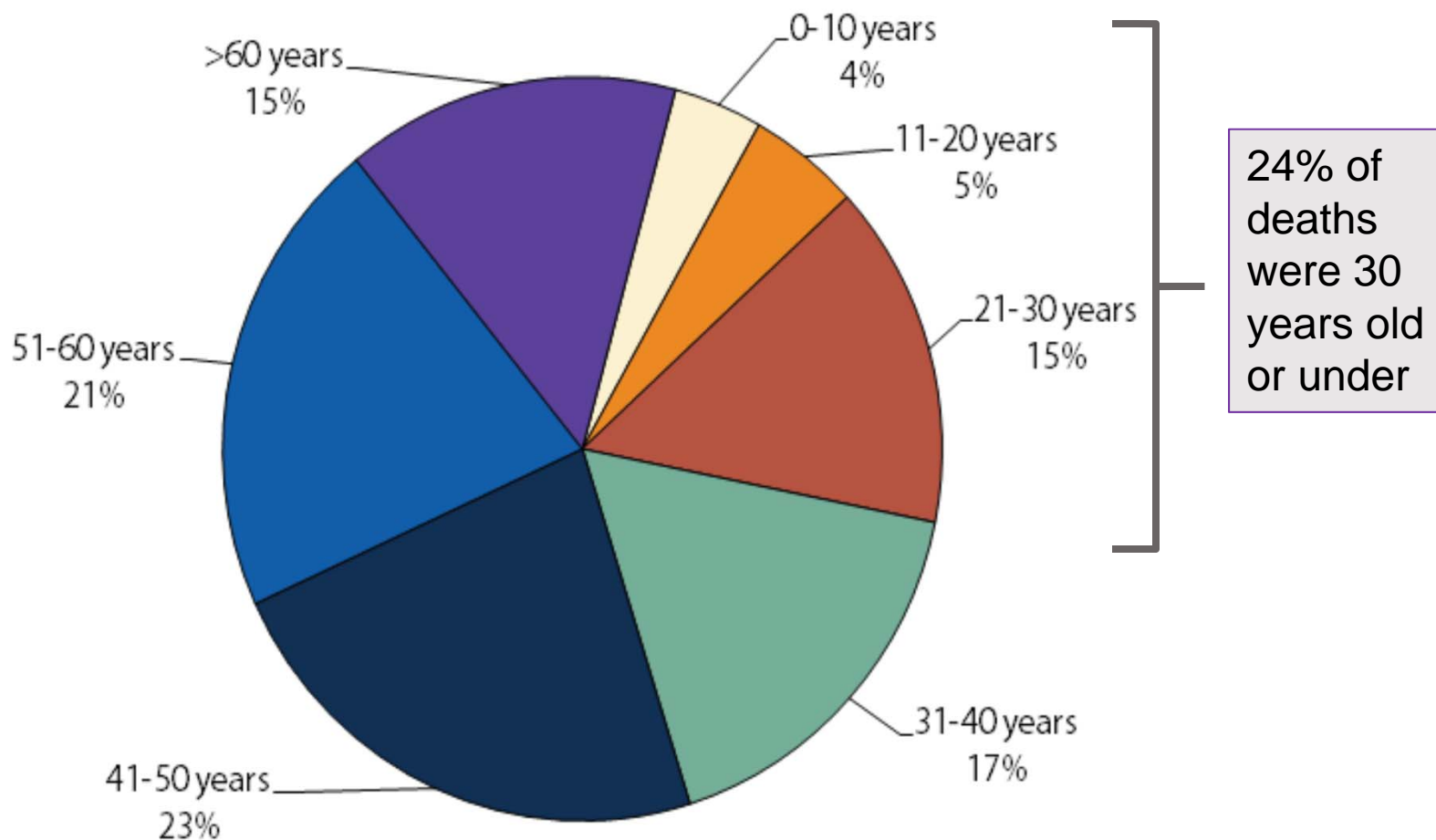


Most common treatments/procedures used among people with SCD on Medi-Cal: 2004-2008



Age at Death for Persons with SCD: 2004-2008

Average age = 43.3 years (n=387)





Case Finding Challenges

- Accessing different data sources
- Difference in case identifiers and data elements across data sources
- Data sources not all easily conducive to research (Medical claims data)
- Developing de-duplication algorithms
- Developing linking algorithms

California Sickle Cell Disease Fact Sheet

Sickle Cell Disease in California

There were approximately 6,200 people with SCD living in California in 2004-2008:

- 43% younger than 18 years
- 21% 18-29 years
- 25% 30-50 years
- 11% 51 years and older

There were 486 babies born with SCD in California in 2004-2008:

- 89% were Black
- 8% were Hispanic
- 5% were Other

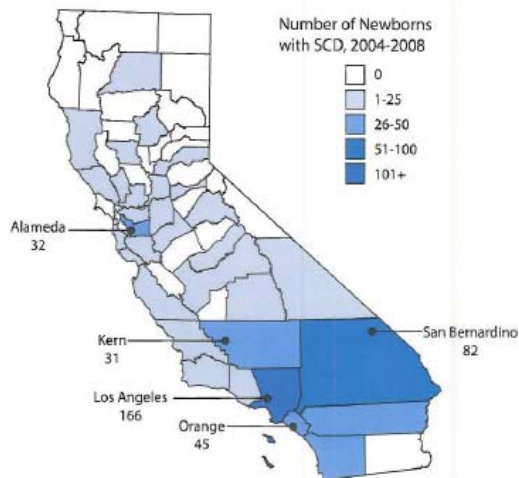
Note: There is overlap between the race and ethnicity categories.

SCD affects all races and ethnic groups. In California, SCD occurs among approximately 1 out of every

- 8,000 births
- 500 Black, African-American, or African descent births
- 99,000 Hispanic-American births

What is Sickle Cell Disease (SCD)?

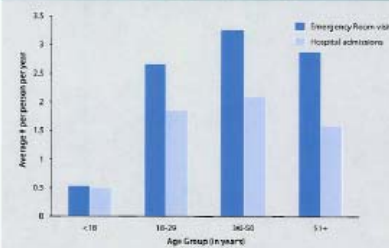
- SCD is a group of inherited conditions that affect hemoglobin, a protein that allows red blood cells (RBC) to carry oxygen to all parts of the body.
- The most common types of SCD are:
 - *Hemoglobin SS Disease (HbSS)* - People who have this form of SCD inherit two sickle cell hemoglobin genes ("S"), one from each parent. This is commonly called *sickle cell anemia* and is usually the most severe form of the disease.
 - *Hemoglobin SC Disease (HbSC)* - People who have this form of SCD inherit a sickle cell hemoglobin gene ("S") from one parent and from the other parent a gene for abnormal hemoglobin called "C". This is usually a milder form of SCD.
 - *Hemoglobin S beta thalassemia (HbS beta thalassemia)* - People who have this form of SCD inherit one sickle cell hemoglobin gene ("S") from one parent and one gene for beta thalassemia, another type of anemia, from the other parent. There are two types of beta thalassemia: "0" and "+". Those with HbS beta⁰-thalassemia usually have a more severe form of SCD. People with HbS beta⁺-thalassemia tend to have a milder form of SCD.



National Center on Birth Defects and Developmental Disabilities
Division of Blood Disorders

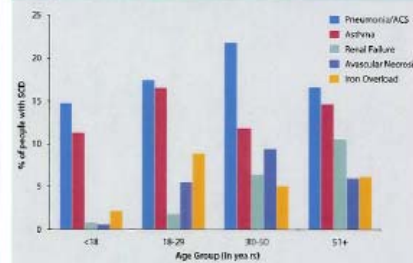


Healthcare utilization by people with SCD, 2004-2008



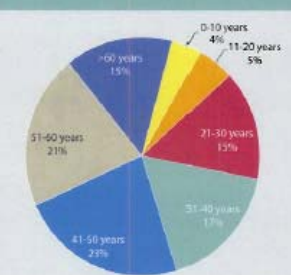
- The number of hospital admissions and emergency room visits rises dramatically after age 18

Most common complications among people with SCD on Medi-Cal, 2004-2008



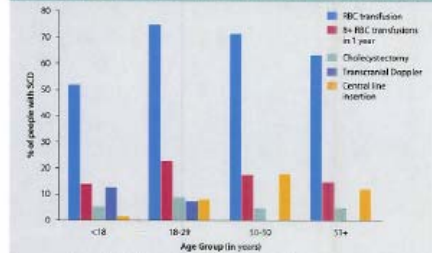
- Pneumonia/Acute Chest Syndrome (ACS) was the most common complication across all ages

Age at death for people with SCD, 2004-2008



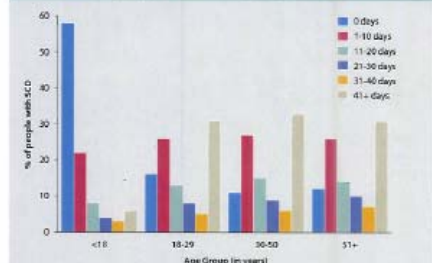
This data was collected through the Registry and Surveillance System for Hemoglobinopathies (RuSH). RuSH was a pilot project that was implemented by the Centers for Disease Control and Prevention (CDC) in collaboration with the National Institutes of Health (NIH), National Heart, Lung, and Blood Institute (NHLBI).

Most common medical treatments and procedures provided to people with SCD on Medi-Cal, 2004-2008



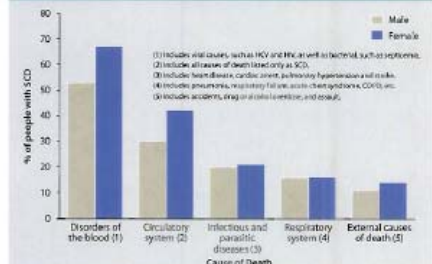
- Over 50% of people with SCD in each age group received at least one transfusion during the five years

Total number of hospital days for people with SCD, 2004-2008



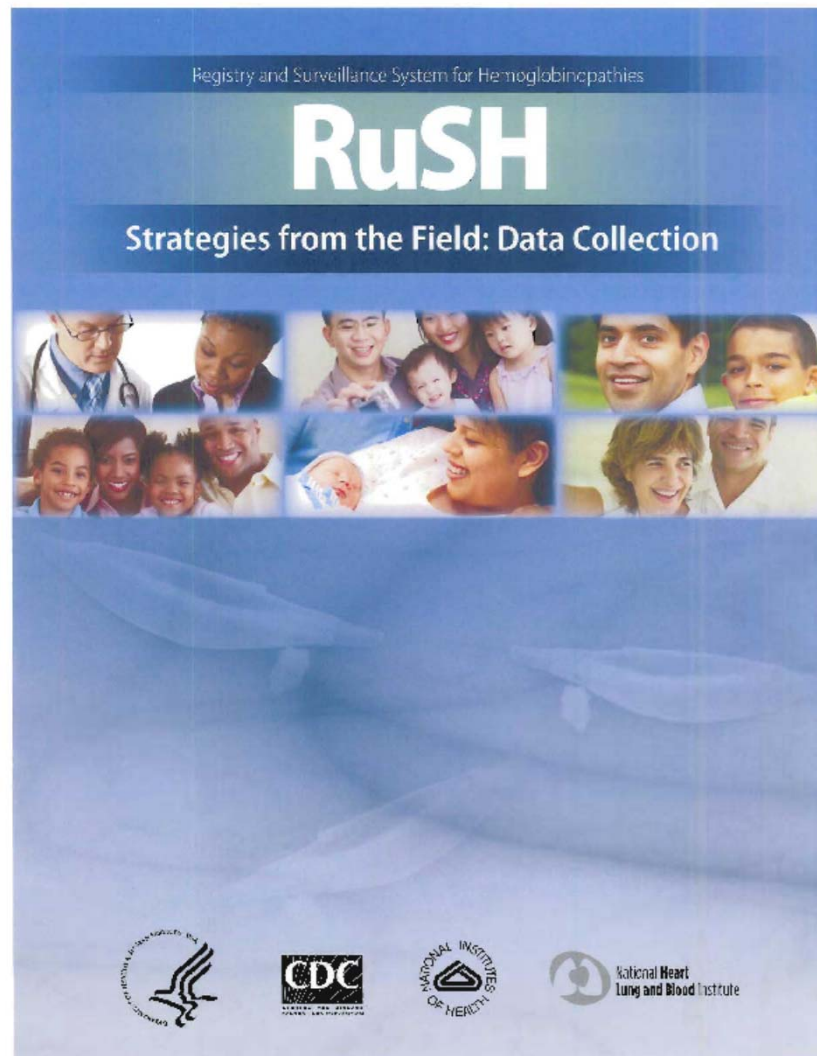
- Over 65% of people with SCD spent at least 1 day in the hospital during the five years

Top 5 causes of death among people with SCD, 2004-2008



For more information, please visit www.cdc.gov/ncbddd/sicklecell and <http://casicklecell.org/>
Like us on Facebook at California Sickle Cell Resources

Strategies From the Field: Data Collection



Next Steps.....



- **Public Health Research, Epidemiology & Surveillance in Hemoglobinopathies – PHRESH**
 - Funded by CDC
 - Summarize and distribute the data collected in RuSH
 - October 2013 through September 2014
 - Validate the RuSH methodology (how well did we do?)
 - Develop disease awareness raising activities
 - Develop health promotion messages for SCD and Thalassemia in California



Thank you!

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[California Sickle Cell Resources](#)

Or at our website at:

CASickleCell.org