National Center on Birth Defects and Developmental Disabilities

DD19-1906 Capacity Building for Sickle Cell Disease Surveillance

Session 11: The SCDC Case Definition

May 21, 2020

SCDC

SICKLE CELL DATA COLLECTION

SCDC Case Definition

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RuSH History

- **RuSH Goal:** Identify and collect data on all people with a **hemoglobinopathy** diagnosis living in the participating states during 2004-2008.
- Three workgroups: The Surveillance Design Work Group **developed case definitions, provided guidance on the interpretation and use of clinical and laboratory information** and identified and refined the clinical variables to be collected and analyzed in the surveillance system.
- Established a three-level case definition for SCD, based on laboratory results and International Classification of Diseases (both the ninth and tenth revisions) coding found in the administrative billing data.
  - Levels were based on the reliability of the diagnosis, with level 1 (confirmed) being the most reliable and level 3 the least (possible).

Original Case Definition

Level 1: Confirmed

- CLIA-certified laboratory result of SCD* reported by a state newborn screening program with confirmatory testing, ***OR*** Clinical diagnosis by a physician with documented confirmatory CLIA-certified laboratory testing after the newborn period

Level 2: Probable

- CLIA-certified laboratory result of SCD reported by a state newborn screening program without report of confirmatory testing, ***OR*** SCD ICD code at two or more separate health-care encounters ***PLUS*** one or more SCD-associated complication, treatment, or procedure

Level 3: Possible

- Sickle cell trait ICD code at two or more separate health-care encounters ***PLUS*** one or more SCD-associated complication, treatment, or procedure ***OR*** SCD ICD code for a single health-care encounter

*Includes hemoglobin S/S, hemoglobin, S/β^0 thalassemia, hemoglobin S/C, S/β^+ thalassemia, and other compound heterozygous forms of SCD.

SCD-Associated Conditions

SCD-associated treatments	SCD-associated complications
Hydroxyurea	Chronic renal failure/proteinuria
Parenteral analgesics (morphine, meperidine, hydromorphone, ketorolac, butorphanol)	Pneumonia, acute chest syndrome
Iron Chelators (deferasirox, deferoxamine)	Pulmonary hypertension
Erythropoietin	Stroke (ischemic or hemorrhagic), transient ischemic attack, seizures
Folic acid	Intracranial bleeding
	Priapism
	Iron overload
SCD-associated procedures	Gallstones/cholelithiasis, cholecystitis
Red cell transfusion	Avascular necrosis
Red cell exchange	Retinal disease
Splenectomy	Splenomegaly, splenic sequestration, hypersplenism
Cholecystectomy	Leg ulcers
Transcranial Doppler	Dactylitis
	Osteomyelitis

Research

PUBLIC HEALTH REPORTS

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SAGE

Improving an Administrative Case Definition for Longitudinal Surveillance of Sickle Cell Disease

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Abstract

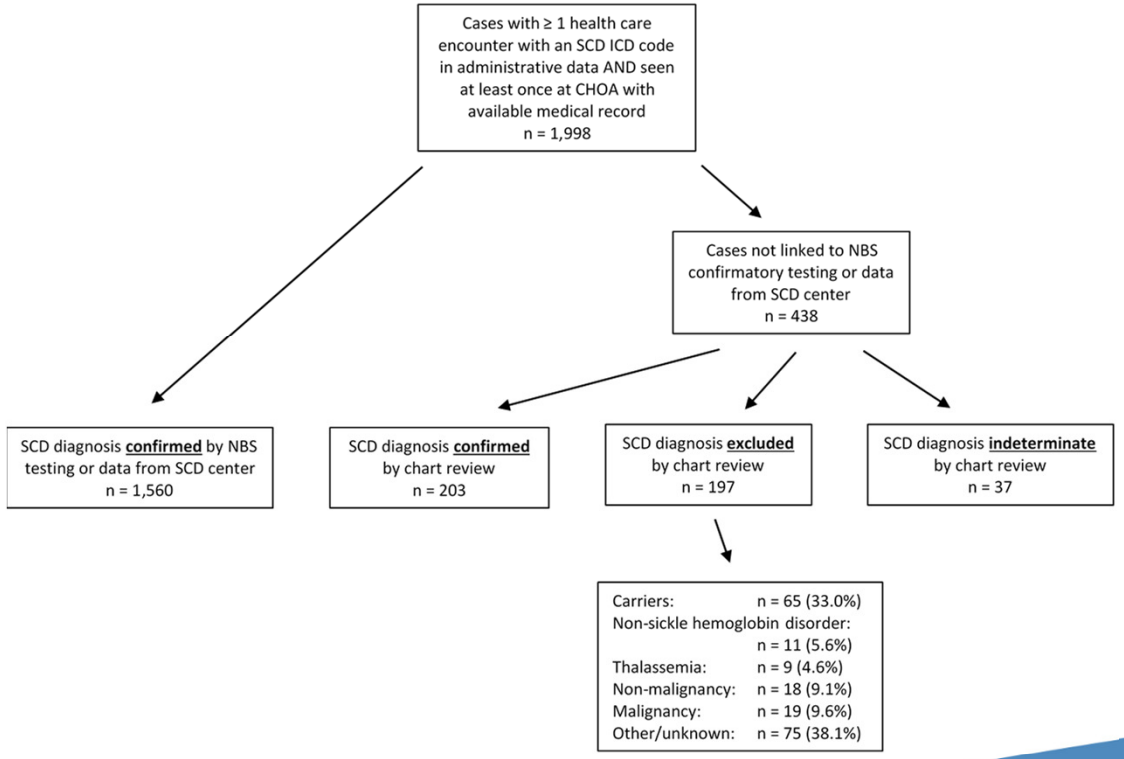
Objective: Several states are building infrastructure and data collection methods for longitudinal, population-based surveillance systems for selected hemoglobinopathies. The objective of our study was to improve an administrative case definition for sickle cell disease (SCD) to aid in longitudinal surveillance.

Methods: We collected data from 3 administrative data sets (2004-2008) on 1998 patients aged 0-21 in Georgia who had ≥1 encounter in which an SCD *International Classification of Diseases, Ninth Revision, Clinical Modification* (ICD-9-CM) code was recorded, and we compared these data with data from a laboratory and medical record review. We assessed performance (sensitivity, specificity, positive predictive value [PPV], and negative predictive value [NPV]) of case definitions that differed by number and type of SCD-coded encounters; addition of SCD-associated treatments, procedures, and complications; and length of surveillance (1 vs 5 years). We identified correct diagnoses for patients who were incorrectly coded as having SCD.

Results: The SCD case definition of ≥3 SCD-coded encounters in 5 years simplified and substantially improved the sensitivity (96.0% vs 85.8%) and NPV (68.2% vs 38.2%) of the original administrative case definition developed for 5-year, state-based surveillance (≥2 encounters in 5 years and ≥1 encounter for an SCD-related treatment, procedure, or complication), while maintaining a similar PPV (97.4% vs 97.4%) and specificity (76.5% vs 79.0%).

Conclusions: This study supports an administrative case definition that specifies ≥3 ICD-9-CM–coded encounters to identify SCD with a high degree of accuracy in pediatric patients. This case definition can be used to help establish longitudinal SCD surveillance systems.

Figure 2 Process Flow to Determine Case Status of Validation Cohort



Performance Metrics for Administrative Case Definitions

Surveillance Period	SCD Case Definition	SCD cases identified	SCD confirmed	SCD cases missed	Non-SCD exclusions	Sensitivity CI**	Specificity CI**	PPV CI**	NPV CI**
5 years	≥ 1 SCD ICD-9 code	1,959	1,763	0	196			90.0%	
5 years	≥ 2 SCD ICD-9 codes	1,831	1,735	28 (1%)	100	98.4% (97.7,98.9)	51.0% (43.8,58.2)	94.8% (93.6,95.7)	78.1% (70.0, 85.0)
5 years	RuSH*	1,558	1,512	251 (14%)	155	85.8% (84.0, 87.4)	79.0% (72.7, 84.6)	97.4% (96.4, 98.1)	38.2% (33.4, 43.1)
5 years	≥ 3 SCD ICD-9 codes	1,739	1,693	70 (4%)	150	96.0% (95.0,96.9)	76.5% (70.0,82.3)	97.4% (96.5,98.1)	68.2% (61.6,74.3)
5 years	≥ 4 SCD ICD-9 codes	1,678	1,647	116 (7%)	165	93.4% (92.2,94.5)	84.2% (78.3,89.0)	98.2% (97.4,98.7)	58.7% (52.7,64.5)
5 years	≥ 5 SCD ICD-9 codes	1,632	1,612	151 (9%)	176	91.4% (90.0,92.7)	89.8.5% (84.7,93.7)	98.8% (98.1,99.3)	53.8% (48.3,59.3)

Optimal Definition

- There is always a trade-off between sensitivity and specificity; depends on the purpose of the study.
- To define the burden of SCD disease for the purposes of public health resource allocation and planning, the number of cases identified should be maximized by choosing a definition that optimizes sensitivity.
- Conversely, for quality and outcome studies of SCD interventions, it is important to limit the number of non-SCD patients in the study sample by maximizing specificity.

SCDC Case Definition

LEVEL 1:

- CLIA-certified laboratory result of SCD reported by a state newborn screening program with confirmatory testing, ***OR*** Clinical diagnosis by a physician with documented confirmatory CLIA-certified laboratory testing after the newborn period

LEVEL 2:

- CLIA-certified laboratory result of SCD reported by a state newborn screening program without report of confirmatory testing, ***OR*** SCD ICD code on three or more separate health-care encounters during a five year period.

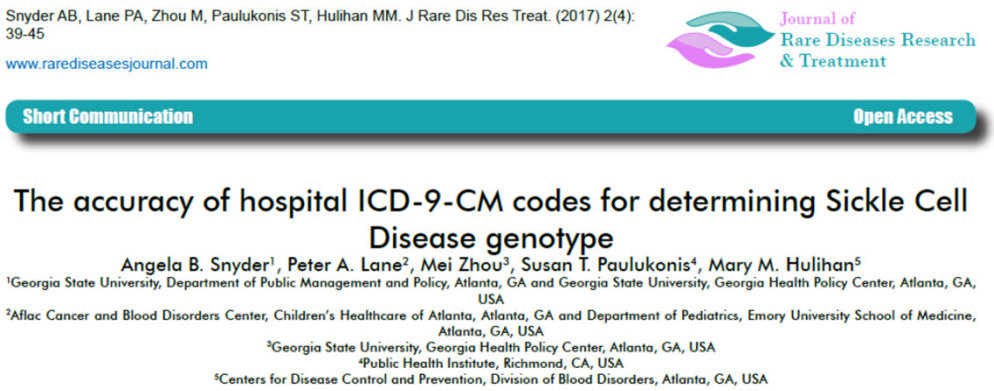
*Includes hemoglobin S/S, hemoglobin, S/β^0 thalassemia, hemoglobin S/C, S/β^+ thalassemia, and other compound heterozygous forms of SCD.

SCD Cases Georgia: 2004-2008

	Confirmed	Probable (RuSH definition)	Probable (SCDC definition)
Newborn Screening	*730*	*98*	*98*
Augusta University (clinical)	*1,218*	*14*	*14*
Grady (clinical)	*1,661*	*2*	*2*
CHOA (clinical)	*1,908*	*242*	*266*
Medicaid/CHIP	*2,983*	*1,987*	*2,079*
State Health Benefit Plan	*209*	*215*	*211*
Hospital discharge	*3,342*	*2,144*	*2,475*
De-duplicated Total	*4,228*	*3,008*	*3,298*

Additional Validation Studies

- Confirmed that using ICD codes to determine SCD genotype is problematic--Sickle-cell/hemoglobin-SS disease seems to be overly represented in hospital reported administrative data.



Snyder AB, Lane PA, Zhou M, Paulukonis ST, Hulihan MM. J Rare Dis Res Treat. (2017) 2(4): 39-45

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Short Communication Open Access

The accuracy of hospital ICD-9-CM codes for determining Sickle Cell Disease genotype

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Case Definition vs Inclusion Criteria

California Sickle Cell Data Collection Program

TRACKING CALIFORNIA

INFORMING ACTION FOR HEALTHIER COMMUNITIES

Standardizing the Population of Interest

- Case definition – how do we standardize who is in our cohort?
  - In the state reporting
  - Diagnosis or combination of diagnostic codes
- Inclusion criteria – how do we standardize who is in this analysis?
  - In the cohort
  - Demographic characteristics (age, sex)
  - Follow-up time frame/length
  - Payer
  - Certainty of diagnostic information
  - Genotype or phenotype (severe/less severe)
  - Geography
  - Specific treatments
  - Deceased/not deceased
  - Seen by knowledgeable care provider
  - Seen in acute care setting
  - Specific comorbidity or condition

CA SCDC (Current) 2016-2018 Cohort

Age Group	Total	In Medi-Cal/CCS	Not in Medi-Cal/CCS
0-9 years in 2018	604	570	34
10-19 years	920	833	87
20-29 years	1,262	1,038	224
30-39 years	1,168	928	240
40-49 years	758	593	165
50-59 years	670	488	182
60-69 years	330	231	99
70-79 years	137	69	68
80 or older	50	21	29
Total	5,899	4,771	1,128

Meeting confirmed or probable case definition with utilization in 2016-2018.

Kayle/Medicaid expansion example

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WILEY

HEMATOLOGY: RESEARCH ARTICLE

Impact of Medicaid expansion on access and healthcare among individuals with sickle cell disease

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Kayle inclusion criteria

Individuals with SCD were included in this study if they were (1) enrolled in Medi-Cal or other state-run programs at any time between 2011 and 2016 for six or more total calendar months in the year and (2) were ≤64 years of age at the close of 2016. The period 2011-2016

TABLE 1 Cohort demographic characteristics

N = 3635	n	%
Age group (years)		
≤11	476	13.1
12-21	680	18.7
22-34	1145	31.5
35-64	1334	36.7
Sex		
Female	2039	56.1
Male	1596	43.9

Johnston/End of life example

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Original Article

Acute Care Utilization at End of Life in Sickle Cell Disease: Highlighting the Need for a Palliative Approach

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Johnston inclusion criteria

Cases. Deceased cohort cases comprised people with SCD who died between 2006 and 2015. Individuals were excluded if their cause of death was due to trauma or peripartum events (either as coded during a terminal admission or on the death certificate) or if no record linkage between the vital records could be made. We adopted this

Characteristic	*Cases* (N = *486*), N (%)
Age group	
0–21 years	41 (8.4)
22–40 years	153 (31.5)
41 years old or older	292 (60.1)
Gender	
Female	259 (53.3)
Male	227 (46.7)

State funded SCD clinics example

- Siting of five new clinics in California to provide care for adults with SCD on Medicaid
- Networking Californians for Sickle Cell Care
- Where are clinics most needed? Where are the sites that are viable for increased capacity that are also needed?
- Criteria:
  - In cohort
  - Acute care utilization 2016-2018
  - At time of acute care utilization, patient zip was within 30 or 60 mile radius of potential location

State funded SCD clinics example

Hospital-Radius	Total Patients that were hospitalized in 2016-2018	Total Patients that had an ED visit in 2016-2018
Big County - 30 mile radius	1071	1449
Big County - 60 mile radius	1630	2162
Antelope Valley - 30 mile radius	121	176
Antelope Valley - 60 mile radius	1173	1631
Kern Medical Center - 30 mile radius	68	84
Kern Medical Center - 60 mile radius	95	125
UCSD Hillcrest Medical Center - 30 mile radius	171	211
UCSD Hillcrest Medical Center - 60 mile radius	199	252
SACHS - 30 mile radius	443	553
SACHS - 60 mile radius	1196	1581
Riverside University Health System- 30 mile radius	426	535
Riverside University Health System - 60 mile radius	912	1184
Loma Linda University Medical Center - 30 mile radius	422	531
Loma Linda University Medical Center - 60 mile radius	1290	1707
Arrowhead Regional Med Ctr. - 30 mile radius	437	542
Arrowhead Regional Med Ctr. - 60 mile radius	1622	2153

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