

Hospitalization and Case Fatality in Individuals with Sickle Cell Disease and COVID-19 Infection



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DISCLOSURES

- This work was supported by Grant 2020079 from the Doris Duke Charitable Foundation
- LM: NIH/NHLBI: Research Funding; NIH/NINDS: Research Funding
- AB: NIH / NHLBI: Research Funding; NIH/NINDS: Research funding; Greater Milwaukee Foundation: Research Funding
- JP: HRSA: Research Funding; NINDS: Research Funding; NINDS: Research Funding; NHLBI: Research Funding

BACKGROUND

- 100,000 individuals with sickle cell disease (SCD) in the United States
- Average life expectancy estimates of 45-55 years
- Respiratory viral infections are source of morbidity and mortality among persons with SCD
- Persons with SCD are more likely to be hospitalized due to influenza infection than the general population
- Importance of understanding impact of new SARS-CoV-2 virus on individuals with SCD

AIMS

- To determine hospitalization and case fatality rates secondary to COVID-19 among individuals living with SCD in different age groups and compare these to the general Black population
 - Hypothesis: SCD patients will have higher hospitalization and case fatality rates compared to general Black population
- To identify factors associated with more severe COVID-19 illness and hospitalization in individuals with SCD
 - Hypothesis: SCD-related comorbidities will be associated with more severe COVID-19 illness and hospitalization

METHODS-Data Source



- Voluntary reporting system
- De-identified data
- Patient demographics
- SCD medical history and comorbidities
- COVID-19 severity, complications and outcome
- Management strategies and healthcare utilization
- <https://covidsicklecell.org/>



METHODS-Data Source

- Case fatality comparison - California Department of Public Health
 - Breakdown of cases and deaths by age and race
 - Publicly available

- Hospitalization comparison - COVID-NET
 - Population-based surveillance system that collects data on laboratory-confirmed COVID-19-associated hospitalizations
 - Network of over 250 acute care hospitals in 14 states
 - Breakdown of hospitalizations by age and race
 - Publicly available

METHODS-Statistical Analyses

- Descriptive statistics
- Indirect age adjustment to calculate standardized mortality ratios (SMR) using COVID-19 data from California state as the reference population
- Multivariable models
 - To assess factors associated with 1) severe COVID-19 infection and 2) hospitalization for COVID-19
 - Age, sex, genotype, hydroxyurea use, SCD-related and non-related comorbidities grouped by organ systems

RESULTS-Study population

Variable	Children (≤ 18) n=152 (41.5%)	Adult (>18) n=214 (58.5%)
Female	71 (46.7)	122 (58.1)
Age, mean (SD)	10.9 (5.4)	34.0 (12.1)
Genotype, HbSS/HbS β 0	109 (71.7)	156 (72.0)
Hospitalization	70 (46.1)	143 (66.8)
Death	1 (0.7)	15 (7.0)

Demographics and characteristics of 366 reported cases in SECURE-SCD Registry, 324 from the United States

RESULTS-Case fatality rate

Age	CDPH cases (n)	CDPH death (n)	Rate (%)	SECURE-SCD cases (n)	SECURE SCD death (n)	Rate (%)
0-17	1998	0	0	127	1	0.8
18-34	8854	35	0.4	116	3	2.6
35-49	6205	64	1.0	59	7	11.9
50-64	6209	238	3.8	17	3	17.6
65-79	3229	483	15.0	5	1	20
80+	1426	489	34.3	0	0	0

Case fatality rate of US cases in SECURE-SCD Registry compared with general Black population from California Department of Public Health data (CDPH)

RESULTS-Hospitalization rates

Age	COVID-NET hospitalized cases (n)	COVID-NET (%)	SECURE-SCD hospitalized cases (n)	SECURE-SCD (%)
0-4	111	0.6	8	4.2
5-17	201	1	50	26.2
18-49	5755	29.5	114	59.7
50-64	6141	31.5	14	7.3
65+	7306	37.4	5	2.6

Hospitalization rate of US cases in SECURE-SCD Registry compared with general Black population from COVID-NET data

RESULTS-Multivariable model

Factors associated with hospitalization in children with SCD

Variable	OR	95% CI		p
Acute care for pain	4.641	1.976	10.9	0.006
SCD Heart/Lung	4.173	1.56	11.158	0.004
Neurobehavioral disorders	2.736	1.099	6.813	0.03

Factors associated with more severe course of COVID-19 illness in children with SCD

Variable	OR	95% CI		p
Acute care for pain	3.964	1.42	11.065	0.03

- **Acute care for pain**
 - 3 or more ED visits or hospitalizations due to acute pain event in last 3 years
- **SCD Heart/Lung**
 - Pulmonary hypertension
 - Acute chest syndrome
- **Neurobehavioral disorders**
 - Anxiety, depression and/or behavioral problems
 - Attention deficit/hyperactivity disorder
 - Developmental delay or learning disability
 - Headaches
 - Seizure disorder
 - Sleep disturbances
 - Stroke

RESULTS-Multivariable model

- Genotype (HbSS/HbS β 0 vs. HbSC/HbS β +) did not show any effect on COVID-19 severity ($p=0.68$) and hospitalization ($p=0.41$) in SCD patients
- Use of hydroxyurea did not show any effect on COVID-19 severity ($p=0.41$) and hospitalization ($p=0.75$) in SCD patients

LIMITATIONS

- Possible selection bias
 - Overrepresentation of more severe patients
 - Underrepresentation of areas where testing is not available
- Case fatality is compared to population of only one state
- No information on date of testing – no time-trend analysis

CONCLUSION

- Individuals with SCD who have COVID-19 infection have higher rates of death due to COVID-19 than the general Black population from California
- Young adults and children with SCD are at higher risk for hospitalization than the general Black population from COVID-NET
- Previous acute care for pain, SCD-related heart and lung comorbidities and neurobehavioral disorders are associated with COVID-19 hospitalization in children with SCD
- Previous acute care for pain is also associated with more severe course of COVID-19 infection in children with SCD