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Characterisation of medical conditions of children with sickle cell disease in the USA: findings from the 2007–2018 National Health Interview Survey (NHIS)

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ABSTRACT

Objectives We used the National Health Interview Survey (NHIS) data set to examine the prevalence of comorbid medical conditions; explore barriers to accessing healthcare and special educational services; and assess the associations between sickle cell disease (SCD) status and demographics/socioeconomic status (SES), and social determinants of health (SDoH) on comorbidities among children in the USA.

Design Cross-sectional.

Setting NHIS Sample Child Core questionnaire 2007–2018 data set.

Participants 133 481 children; presence of SCD was determined by an affirmative response from the adult or guardian of the child.

Main outcome measures Multivariate logistic regression was used to compare the associations between SCD status, SES and SDoH for various medical conditions for all races and separately for black children at p<0.05.

Results 133 481 children (mean age 8.5 years, SD: 0.02), 215 had SCD and ~82% (weighted) of the children with SCD are black. Children with SCD were more likely to suffer from comorbid conditions, that is, anaemia (adjusted OR: 27.1, p<0.001). Furthermore, children with SCD had at least two or more emergency room (ER) visits (p<0.001) and were more likely to have seen a doctor 1–15 times per year (p<0.05) compared with children without SCD. Household income (p<0.001) and maternal education were lower for children with SCD compared with children without SCD (52.4% vs 63.5% (p<0.05)). SCD children with a maternal parent who has < / > High School degree were less likely to have no ER visits or 4–5 ER visits, and more likely to have 2–3 ER visits within 12 months.

Conclusion Children with SCD experienced significant comorbid conditions and have high healthcare usage, with black children being disproportionately affected. Moreover, maternal education status and poverty level illustrates how impactful SES can be on healthcare seeking behaviour for the SCD population. SDoH have significant implications for managing paediatric patients with SCD in clinical settings.

STRENGTHS AND LIMITATIONS OF THIS STUDY

⇒ This study uses a robust and representative data set, which allows for an accurate measure of the impact of sickle cell disease (SCD), a rare condition, and the effect of socioeconomic status (SES) on medical comorbidities and on social determinants of health domain of healthcare usage for this population.

⇒ National Health Interview Survey (NHIS) contains data for children of various races, SES, insurance coverage and geographical regions throughout the USA. This diversity and range of information makes it possible to compare children based on SCD status and substantiates the generalisability of our findings.

⇒ The survey weights provided by NHIS were used to weigh the sample to prevent skewing from categories with smaller sample sizes.

⇒ Because SCD was of a low prevalence in this population, it is possible that selection bias influenced some of the associations examined in this secondary analysis, mainly because we compared a small number of children with a condition that is not well represented in the general population of children across the USA.

INTRODUCTION

Sickle cell disease (SCD) is a group of inherited red blood cell disorders characterised by abnormal sickle-shaped erythrocytes in the blood.[1] SCD is one of the most common genetic disorders in the USA, affecting 70 000–100 000 children and adults of predominantly African descent.[2] Among blacks, SCD is diagnosed in 1 of 365 births.[3] Children with SCD have been observed to have a lower health-related quality of life due to the impact of severe disease-related complications and comorbidity.[4] These complications

and comorbidity include infections, pain crisis,[5–8] acute chest syndrome, metabolic deficiencies,[9] overt stroke[4] and even educational challenges that begin in elementary school and persist post high school,[10 11] in the presence of other comorbid conditions.

Existing evidence suggests a significant increase from 2006 to 2015 in direct hospital admissions of children with SCD.[1] Emergency department-based admissions account for 80% of all medical expenditures, which includes services received by patients with SCD.[1 12] The current literature also suggests that children with SCD predominantly belong to families of lower socioeconomic status (SES) and parents with lower educational attainment,[12] a social determinant of health (SDoH).[13] Furthermore, studies have shown considerably higher healthcare usage among children with SCD from low SES families, whose SES status directly affects the timeliness and quality of treatment.[12] Additional documented barriers to care include lack of access to healthcare facilities, lack of knowledge about SCD, limited SCD management options; or denial of care by specialised health facilities due to having a healthcare plan with lower reimbursement rates.[1] Psychosocial factors such as stigma,[14 15] previous negative healthcare experiences and poor healthcare satisfaction have also been associated with the delay of care.[14] Yet, limited studies are using population-level data to document SCD-related comorbidities and complications and the additional burden on children and their families.

Studies using US population-based data to characterise the health status and healthcare usage patterns are scant. Except for the study conducted by Boulet *et al* in 2010, which examined the 1997–2005 data from the National Health Interview Survey (NHIS), an annual, multistage probability sample survey from a non-institutionalised representative population in the USA to describe health status and health services use among black children aged 0–17 years with SCD.[16] Findings from Boulet *et al* study indicate that black children with SCD (n=192) were more likely to report delays in accessing healthcare compared with those without SCD (n=19 335), thus highlighting the existing barriers to medical care for the SCD population. The authors emphasised the need for additional studies examining the extent of the healthcare access dilemma for US children with SCD from low SES families. Our study builds on this very informative work. We used data from the 2007–2018 NHIS to describe the health status and health services use among children of all races/ethnicities aged 0–17 years with SCD.[17] Specifically, we examined the (1) prevalence of comorbid medical conditions; (2) the usage and barriers to accessing healthcare and special educational services; and (3) assess the associations between SCD status and demographics/SES on comorbid medical conditions and healthcare usage. We hypothesised that black children with SCD would have higher levels of disease-related complications and comorbidities, as well as healthcare usage than those without SCD.

METHODS

Study design

Secondary analysis of NHIS 2007–2018 data set

This cross-sectional study used data reported in the Sample Child Core questionnaire portion of the NHIS survey (2007–2018).[18] NHIS data comprises a range of health topics that provide data to track health status, healthcare access and progress toward achieving national health objectives. In each of the years, an individual between the age of 0–17 years old was randomly selected from participating households to participate as a subject for the Sample Child Core questionnaire. In addition, a parent or legal guardian knowledgeable about the child's medical conditions and healthcare needs was selected to provide a proxy response on behalf of the sampled child.

Data collection

NHIS data was collected via an in-person interview. Parents of the 133 481 children (final study sample) were asked about SCD diagnosis or status of their children. This study included children aged 0–17 who participated in the Sample Child Core questionnaire part of the NHIS from 2007 to 2018. The initial unweighted sample included 133 542 children, with 68 745 male children and 64 797 female children.

Survey measures

The Sample Child Core questionnaire inquiries about a child's health status, functional limitations and access to healthcare. The presence of SCD was determined by an affirmative response from a parent/guardian representing the child as such: 'Looking at this list, has a doctor or other health professional ever told you that [SC Child] had any of these conditions? Which ones… sickle cell anaemia,…, etc.?' Medical insurance information was collected via questionnaires and classified into three categories: public, private or other. The household income level was defined according to the federal poverty level, and we classified these into three categories: <100%, 100% to <200% and ≥200% of the federal poverty level.[19] The annual US poverty level for an individual was defined as income of US$10 210 (2007) and US$12 140 (2008).[20 21] The US poverty rate during the initial period of study (2007) was 12.5% (total), and 24.5%, 10.5% for blacks and whites, respectively.[22] In 2018, the overall poverty rate was 11.8%; and the rate for blacks (20.8%) remain twice that of whites (10.1%).[23]

Health conditions were obtained via questions about participants' medical history. Health impact was evaluated, which assessed functional status (walking, playing, etc), taking prescription medication for at least 3 months, health status compared with 12 months ago and school absences due to illness/injury. Health status was classified into three categories: better, worse and about the same. Duration for missing school due to illness was divided into six categories: none, single days, more than 10 days, more than 20 days, more than 30 days and more than 40 days. Healthcare usage, an SDoH was assessed via

questions about accessing medical services including specialists, psychiatrists, psychologists, psychiatric nurses, clinical social workers, physical therapists, speech therapists, respiratory therapists, audiologists, occupational therapists, optometrists, ophthalmologists or any surgical procedures. Frequency of annual physician visits was stratified into five categories: none, 1–5 times, 6–9 times, 10–15 times and 16 or more times. Frequency of annual emergency room (ER) visits were stratified into six categories: none, 1 time, 2–3 times, 4–5 times, 6–16 or more times and underwent surgical procedure.

Healthcare barriers were assessed using the following question, 'Have you delayed getting care for {S.C. name} for any of the following reasons in the PAST 12 MONTHS?' Response options included the following: challenges getting an appointment, affording prescription medicines and difficulty obtaining transportation to the medical service location.

For the comorbid medical conditions queried for this study, respondents were asked whether the following medical conditions had occurred during the 12 months preceding the interview: anaemia, asthma attack, hay fever, respiratory allergy, food or digestive allergy, eczema or skin allergy, frequent diarrhoea or colitis, frequent severe headaches, or migraines and three or more ear infections.

Statistical analyses

Statistical analyses were performed using Stata software (V.16.0).[24] Frequencies and proportions were obtained for all categorical variables and means and SD for continuous variables. All weighted results were calculated to represent national estimates for children between 0 and 17 years old.

We evaluated frequencies and weighted percentages of demographic characteristics among children of all races (table 1) and separately for black children (table 2) by SCD status. We also examined the associations between SCD status and comorbid medical conditions while controlling for demographic characteristics in multivariate logistic regression models among of all races (table 3) and for black children (table 4), and for health status and healthcare use (tables 5 and 6). We then conducted analyses of the various potential interactions between SCD status and demographic characteristics impacting the medical condition/health status/healthcare usage of children using multivariate logistic regression models (online supplemental table 1). We estimated the statistical significance based on p values at a significance level of p<0.05, measured main effects with adjusted ORs (AORs) and interpreted interaction effects with margins plots. The interaction terms were selected from tables 1 and 2. Participants missing SCD question responses (<1%) were excluded from the analyses.

Patient and public involvement

No patients were actively involved in setting the research question, outcome measures nor involved in the design of the study. Patients were not involved in interpretation or write up of the results, nor are there plans for the results to be disseminated to the patient affected by this research.

RESULTS

Characteristics of sample

As presented in table 1, figure 1 and online supplemental figure 1, the sample included 133 481 children (mean age 8.5 years, SD: 0.02) across the 12 years and we identified 215 children with SCD (0.16%). Of the children with SCD, the mean age was 8.2 years (SD: 0.29) and 110 are men. Racial composition among children with SCD includes black (81.8%), multiple and other races (8.7%) and white (7.7%). Of the children with SCD, 49.1% are from the Southern part of the USA. More than half (53.8%) of children who have SCD reported a household income less than 100% of the federal poverty level (online supplemental figure 2). 53.9% of children with SCD reported a Medicaid and/or a State Children's Health Insurance Program (SCHIP) as their primary insurance (figure 2). Of the children without SCD, 52% have private insurance. Among children with SCD, 28% of maternal respondents reported having only a high school education. The subanalysis of black children with SCD showed a similar pattern with 55% reporting a household income <100% of the federal poverty level; and 55.8% receive Medicaid/SCHIP insurance (table 2). Across the 12 years, the period prevalence of SCD among all children was 1.47 per 1000 (SE: 1.4; 95% CI: 1.2 to 1.8). Black children had a higher period prevalence of SCD (7.83 per 1000; SE: 7.94; 95% CI: 6.3 to 9.4) and overall, an increased burden of medical conditions.

Medical conditions, health status and healthcare usage

Among the sample, children with SCD experienced a greater proportion of medical conditions within the past 12 months compared with children without SCD (table 3, online supplemental figure 3). After applying the appropriate weights to each of the demographics, all children with SCD had higher AOR of having anaemia (AOR 27.1; 95% CI: (15.6 to 46.9); p<0.001), respiratory allergies (AOR 2.2; 95% CI: (1.3 to 3.9); p=0.004) and frequent severe headaches/migraines (AOR 2.1; 95% CI: (1.1 to 4.1); p=0.034) (table 3). Furthermore, children with SCD are twice as likely to have seen a medical specialist (AOR 2.3; 95% CI: (1.5 to 3.7); p<0.001), and a therapist (AOR 95% CI: (1.0 to 4.1)) within the past 12 months. Because of the limited number of children with SCD, we could not examine the association between SCD and 'hay fever'.

Compared with black children without SCD, black children with SCD had a higher odd of having anaemia (AOR 18.6; 95% CI: (10.6 to 32.6); p<0.001) and respiratory allergies (AOR 2.4; 95% CI: (1.3 to 4.3); p=0.005) (table 4). Children with SCD are also more likely to experience other comorbid conditions including food/digestive allergies, eczema/skin allergies and frequent severe headaches/migraines compared with children without

Table 1 Demographic characteristics of all children by sickle cell disease (SCD) status (N=133 481)

	Children with SCD (n=215)		Children without SCD (n=133 266)	
	Unweighted n	Weighted %	Unweighted n	Weighted %
Year of survey				
2007	17	8.36	9396	8.33
2008	17	9.85	8796	8.34
2009	18	8.25	11 133	8.36
2010	27	9.77	11 241	8.43
2011	20	7.40	12 826	8.42
2012	27	8.89	13 239	8.32
2013	24	10.39	12 828	8.30
2014	14	7.02	13 361	8.30
2015	15	6.11	12 271	8.30
2016	21	14.01	11 081	8.30
2017	5	2.59	8837	8.31
2018	10	7.35	8257	8.30
Child gender				
Male	110	48.23	68 593	51.07
Female	105	51.77	64 673	48.93
Child age (years)				
<3	42	16.30	23 052	16.23
3–5	45	23.75	21 571	16.99
6–10	49	22.47	34 053	27.71
11–14	37	20.42	29 077	22.15
15–17	42	17.06	25 513	16.92
Race				
White	16	7.66	85 965	68.83
Black/black	170	81.81	21 395	15.34
Indian (American Indian)	1	0.57	1847	1.34
Asian	2	1.22	5458	3.12
Other race	21	6.41	16 705	10.13
Multiple race	5	2.33	1896	1.25
Maternal education				
<High school	40	19.02	19 310	14.81
High school/GED	54	28.58	27 421	21.68
>High school	99	52.41	75 560	63.51
Missing	22		10 975	
Household income (% federal poverty level)				
<100	97	53.81	23 303	21.00
100 to <200	42	22.51	26 787	22.94
≥200	42	23.68	64 984	56.05
Missing	34		18 192	
Insurance				
Medicaid/SCHIP	111	53.85	32 653	25.84
Private insurance	51	27.63	62 976	52.12
Other insurance	18	7.42	10 346	7.92
No insurance	17	7.77	10 405	7.43

Continued

Table 1 Continued

	Children with SCD (n=215)		Children without SCD (n=133 266)	
	Unweighted n	Weighted %	Unweighted n	Weighted %
Multiple insurance	8	3.33	8043	6.69
Missing	10		8843	
Region				
Northeast	30	14.56	20 949	16.27
Midwest	50	30.40	27 091	22.82
South	116	49.07	48 356	36.87
West	19	5.97	36 870	24.04

SCHIP, State Children's Health Insurance Program.

SCD, although statistically non-significant (p>0.05). All children (28.9%) and black children (29.9%) with SCD had at least two or more annual ER visits and were more likely to have seen a doctor more than five times per year than all children and black children without SCD, respectively, (tables 5 and 6). Additionally, all children with SCD were seven times more likely to have limited ability to crawl, walk, run or play, and three times more likely to take prescription medications for at least 3 months compared with all children without SCD (p<0.001).

Parents of children with SCD were four times more likely to report worsening health status over 12 months compared with parents of children without SCD (p<0.01). Additionally, children with SCD were less likely to report the same health status over 12 months compared with children without SCD (p<0.01) (table 5). Compared with children without SCD, children with SCD were more likely to miss 11–40+ days of school per year (p<0.001). Black children with SCD were more likely to miss 11–40+ school days because of illness/injury than black children without SCD (p<0.001) (table 6). In terms of healthcare access barriers, only one black child with SCD could not afford prescription medications as reported in NHIS compared with black children without SCD (table 6).

Association of SCD status and demographics/SES with medical comorbidities with healthcare usage

We examined potential interaction effects between SCD and SES and its impact on medical comorbidity and on SDoH domain of healthcare usage among the sampled children (see online supplemental table 1 and figures 3A–G, 4A–K, 5A–C). Although many interactions exist, in this section, we only discussed statistically significant interaction items (p<0.05) in our study with margin plots.

Considering medical conditions in the past 12 months, the effects between SCD and anaemia depend on race (black: p=0.008). The probability of anaemia is higher for black children than non-black children without SCD, opposite to the results of children with SCD. Additionally, other insurance (any insurance other than Medicaid and private insurance) contributed as an interaction term to the effects between SCD and two types of allergies, which are respiratory allergies (p=0.06) and digestive allergies (p=0.037). The chances of having respiratory and digestive allergies are statistically significant for other insurance when children are without SCD, which is different from the conclusion of children with SCD.

Age (3–5: p=0.04, 11–17: p=0.01) and gender (male: p=0.01) interacted with SCD and influenced the association between SCD and taking prescription medication for at least 3 months. SCD had a more considerable effect among men than women on taking prescription medication. The probability of taking prescription medication for children without SCD ages 11–17 (p=0.001) is the highest among the four age categories; and that for SCD children ages 3–5 (p=0.04) is the lowest.

Maternal education levels, age, region and income had interaction effects with SCD on doctor visits. Compared with children without SCD, children with SCD with a maternal parent were less likely to visit the doctor within 12 months. Interactions between SCD and maternal education as high school/GED (p=0.048) or above (p=0.040) were statistically significant on not seeing a doctor compared with children with SCD with a maternal parent having less than a high school education. Furthermore, certain SES factors interacted with the degree of healthcare usage. For example, children with SCD having 1–5 annual doctor visits were more likely to have a household income between 100% and <200% of the federal poverty level (p=0.046) and live in the Southern region of the USA (p=0.034). Southern region interacted even more as the frequency of annual doctor visits increased to 6–9 visits (p=0.008) and living in the Midwest region also interacted at the same level of usage (p=0.02). Age was an important factor across many of the age groups, with increasing interaction for children with SCD having 6–9 annual doctor visits (3–5: p=0.031, 6–10: p=0.002) and 10–15 annual doctor visits (6–10: p=0.047, 11–17: p=0.046). The possibility of visiting a doctor 1–5 times per year for household income below 100% of the federal poverty level was the lowest among other levels when children were without SCD but the most considerable

Table 2 Demographic characteristics of black children by sickle cell disease (SCD) status (N=21 565)

	Black children with SCD (n=170)		Black children without SCD (n=21 395)	
	Unweighted n	Weighted %	Unweighted n	Weighted %
Year of survey				
2008	16	11.58	1535	8.38
2009	16	9.60	1961	8.45
2010	22	10.48	2000	8.36
2011	19	8.01	2172	8.26
2012	20	8.83	2221	8.12
2013	18	10.68	2148	8.33
2014	9	6.60	2089	8.25
2015	9	3.57	1848	8.28
2016	16	13.00	1455	8.26
2017	3	1.96	1163	8.41
2018	8	6.68	1124	8.21
Child gender				
Male	86	48.71	10 985	50.47
Female	84	51.29	10 410	49.53
Child age (years)				
<3	31	14.37	3459	15.99
3–5	33	23.54	3541	17.55
6–10	38	20.99	5429	27.72
11–14	32	22.36	4794	22.04
15–17	36	18.74	4172	16.70
Maternal education				
<High school	25	15.94	2519	14.26
High school/GED	44	30.28	5057	26.79
>High school	83	53.77	11 243	58.94
Missing	18		2576	
Household income (% federal poverty level)				
<100	78	55.08	5967	36.21
100 to <200	32	24.72	4998	26.89
≥200	30	20.20	7216	36.90
Missing	30		3214	
Insurance				
Medicaid/SCHIP	91	54.84	9285	44.51
Private insurance	42	26.86	7147	34.33
Other insurance	14	7.50	1852	9.65
No insurance	12	8.09	1375	6.29
Multiple insurance	4	2.69	962	5.23
Missing	7		1717	
Region				
Northeast	23	15.54	3422	16.07
Midwest	44	33.89	3747	19.07
South	94	48.67	12 092	55.95
West	9	1.91	2134	8.91

SCHIP, State Children's Health Insurance Program.

Table 3 Medical conditions among all children by sickle cell disease (SCD) status (N=133 481)

	Children with SCD n=(215)		Children without SCD n=(133 266)		AOR for SCD vs no SCD (95% CI)	P value
	Unweighted n	Weighted %	Unweighted n	Weighted %		
Medical conditions (past 12 months)						
Anaemia	54	30.65	1188	1.04	27.1 (15.6 to 46.9)	<0.001
Asthma attack	31	39.10	6619	37.80	0.9 (0.4 to 1.7)	0.709
Hay fever	0	–	675	2.91	–	–
Respiratory allergies	33	18.05	13 006	11.53	2.2 (1.3 to 3.9)	0.004
Food/digestive allergies	16	8.38	6062	5.39	1.8 (0.9 to 3.7)	0.084
Eczema/skin allergy	40	22.06	12 928	11.66	1.6 (0.9 to 2.6)	0.088
Frequent diarrhoea/ colitis	4	1.39	1517	1.36	1.2 (0.4 to 3.5)	0.655
Frequent severe headaches/migraines	26	11.27	6695	5.75	2.1 (1.1 to 4.1)	0.034
Three or more ear infections	8	2.72	4508	4.11	0.6 (0.2 to 1.7)	0.378

AOR, adjusted OR.

among children with SCD. Children without SCD (age <3) had the most significant chance of having 6–9 doctor visits per year but those with SCD of the same age, had the third largest chance of having 6–9 doctor visits. For the ER visit frequency, the demographics/SES, including age, maternal education, region, income and insurance, interactions exist (p<0.05) with SCD. Men (p<0.012), household income (>200: p<0.022) and private insurance (p<0.001) had enormous effects that interacted with SCD for children who had surgery.

DISCUSSIONS

We compared medical conditions, health status and healthcare usage among a nationally representative sample of children with and without SCD. Results from this study suggest that children in the USA who have SCD, regardless of racial/ethnic background, are more likely to suffer from comorbid medical conditions, have greater healthcare usage and experience more health consequences and healthcare barriers than children without SCD. This is consistent with results from the

Table 4 Medical conditions among black children by sickle cell disease (SCD) status (N=21 565)

Medical conditions (past 12 months)	Black children with SCD n=170		Black children without SCD n=(21 395)		AOR for SCD vs no SCD (95% CI)	P value
	Unweighted n	Weighted %	Unweighted n	Weighted %		
Anaemia	42	27.17	312	1.70	18.6 (10.6 to 32.6)	<0.001
Asthma attack	29	44.10	1732	40.69	1.1 (0.5 to 2.4)	0.775
Hay fever	0	–	106	2.77	–	–
Respiratory allergies	28	18.73	2215	11.50	2.4 (1.3 to 4.3)	0.005
Food/digestive allergies	12	7.91	1061	5.85	1.7 (0.8 to 3.7)	0.203
Eczema/skin allergy	33	22.03	2926	16.26	1.4 (0.8 to 2.4)	0.286
Frequent diarrhoea/ colitis	2	0.54	234	1.30	0.4 (0.1 to 2.0)	0.289
Frequent severe headaches/migraines	23	10.29	1282	6.58	1.6 (0.8 to 3.2)	0.191
Three or more ear infections	5	1.82	528	3.07	0.5 (0.1 to 1.8)	0.257

AOR, adjusted OR.

Table 5 Health status and healthcare use for all children by sickle cell disease (SCD) status (N=133 481)

	Children with SCD **Unweighted n, n=(215)**		Children without SCD **Unweighted n, n=(133 266)**		AOR for SCD vs no SCD **(95% CI)**	**P value**
	Unweighted n	**Weighted %**	**Unweighted n**	**Weighted %**		
Health impact						
Limited in ability to crawl, walk, run or play	22	12.70	2340	1.75	7.2 (3.9 to 13.3)	<0.001
Needed special equipment	4	1.47	1566	1.16	1.3 (0.4 to 4.4)	0.647
Took prescription medication for at least 3 months	78	36.44	17 308	12.95	3.8 (2.4 to 6.2)	<0.001
Reported health status fair or poor						
Better	69	33.42	29 475	20.73	1.5 (1.0 to 2.3)	0.035
Worse	10	4.63	1792	1.33	4.1 (1.8 to 9.0)	0.001
About the same	135	61.95	101 648	77.95	0.5 (0.4 to 0.8)	0.003
Miss school because of illness or injury (past 12 months)						
0	29	24.85	28 969	30.12	0.4 (0.2 to 0.8)	0.007
1–10	78	59.37	60 803	65.45	1.1 (0.7 to 1.8)	0.723
11–20	14	8.82	3176	3.26	3.8 (1.8 to 7.7)	<0.001
21–30	5	2.84	746	0.71	6.5 (2.4 to 17.6)	<0.001
31–40	0	–	174	0.15	–	–
40+	5	4.11	328	0.31	14.0 (4.5 to 43.1)	<0.001
Healthcare and special education services use (past 12 months)						
Saw a medical specialist	56	23.56	18 836	14.29	2.3 (1.5 to 3.7)	<0.001
Saw mental health professional	22	9.82	9219	7.81	1.04 (0.5 to 2.2)	0.911
Saw a therapist	22	13.20	8558	7.55	2.0 (1.0 to 4.1)	0.040
Saw an optometrist, ophthalmologist or eye doctor	57	31.20	31 873	26.89	1.4 (0.9 to 2.3)	0.126
How many times have you seen a doctor within 12 months?						
None	9	3.06	12 651	9.19	0.2 (0.1 to 0.6)	0.003
1–5	142	68.47	99 137	75.91	0.5 (0.4 to 0.8)	0.004
6–9	28	14.72	11 842	8.82	2.6 (1.4 to 4.8)	0.002
10–15	21	9.55	5239	3.86	3.5 (1.9 to 6.4)	<0.001
16 or more	12	4.20	2921	2.22	2.2 (0.9 to 5.2)	0.073
Multiple visits to emergency room						
None	125	55.32	106 956	81.24	0.4 (0.2 to 0.6)	<0.001
1	34	15.75	16 969	12.50	1.0 (0.5 to 1.7)	0.935
2–3	32	17.28	6869	5.01	2.9 (1.6 to 5.2)	0.001
4–5	13	6.54	1089	0.79	4.8 (2.3 to 10.2)	<0.001
6–16+	11	5.11	634	0.45	9.3 (3.6 to 24.0)	<0.001
Had surgery or another surgical procedure	15	3.92	6286	4.76	1.0 (0.5 to 2.0)	0.957
Healthcare barrier						
Could not use telephone	5	3.62	2330	1.74	1.4 (0.4 to 5.5)	0.613
Could not get an appointment	11	5.66	5882	4.45	1.1 (0.5 to 2.5)	0.858

Continued

Table 5 Continued

	Children with SCD Unweighted n, n=(215)		Children without SCD Unweighted n, n=(133 266)		AOR for SCD vs no SCD (95% CI)	P value
	Unweighted n	Weighted %	Unweighted n	Weighted %		
Waited too long at the doctor's office	11	2.63	5457	3.95	0.6 (0.2 to 1.3)	0.163
No transportation	12	4.42	2416	1.83	0.8 (0.3 to 1.9)	0.646
Could not afford prescription medicines	1	5.45	230	1.37	5.1 (0.9 to 28.3)	0.065

AOR, adjusted OR.

previous 1997–2005 NHIS analyses.[16] Despite children with SCD having more visits to both doctors' offices and the ER,[25] they reported having more healthcare barriers than children without SCD.[16] We observed a similar trend among black children. For children with SCD, prescription medication affordability poses a significant financial burden for their families, especially among low-income households. Unfortunately, lack of adequate and timely treatment results in deteriorating health, supported by evidence from our study whereby black children with SCD reported worsening health over 12 months. Moreover, based on our findings, maternal education status and poverty level illustrates the impact of SES on healthcare seeking behaviour for the SCD population. Compared with the poverty rates for the general African American US population,[22 23] the poverty level among African Americans without SCD participating in NHIS was higher; and significantly greater for the SCD population, and the majority of the respondents reported Medicaid as their primary insurance. Public insurance may limit accessibility to specialised healthcare services and pose tremendous challenges to receiving timely care.[26] Also, lack of health insurance may result in children settling for ER visits as their sole option for primary care.[27] In this sample, we found that 28.9%–29.9% of children with SCD had at least two or more annual ER visits.

As the current literature suggests, children with SCD typically come from families of a lower SES[12] and therefore, experience significant social challenges when managing their condition, often waiting too long to seek care.[28] Additionally, children with SCD have higher healthcare usage than other low-income children due to the need to seek care for multiple conditions related to their SCD,[12] and also experience more healthcare barriers.[29] Therefore, household SES (ie, parental education),[30–33] can directly affect the frequency of healthcare usage, matching our analyses' trends. Although, visiting ER/urgent care frequently is often necessary for individuals living with SCD due to the nature of the condition and irrespective of their SES,[14 25] existing studies have indicated that patients are hesitant to visit the ER/urgent care due to previous negative healthcare experiences and stigmatisation[14 15 34] because of providers' implicit biases.[35] We acknowledge that there were many conflicting observations within the NHIS data set (eg, both highly educated and less than high school educated mothers visited the emergency department less often). We attribute this to the stigmatisation that many African Americans encounter when seeking healthcare for SCD management. Stigmatisation experiences are prevalent within the African American community despite the level of education due to historical medical maltreatment. However, we do not have enough data based on NHIS to disentangle the contradictory observations, but those observations provide an opportunity for further research and avenues to improve health outcomes and decrease stigma for people living with SCD.

Among our SCD cohort, we observed that black children with SCD reported more visits to the ER perhaps due to disease severity. As done with our study, synthesis of available representative population-level data is an added value to the documentation of the SCD burden among US populations and it is critical for deriving context-appropriate solutions and informed scale-up of effective SCD management therapies including disease-modifying agents (ie, hydroxyurea),[36 37] and nutritional supplements, which have been reported to improve pain crisis and decrease hospitalisations.[38 39] While we examine these patterns of healthcare usage, we are aware that use of prescription medicines and use of subspecialist visits may not necessarily be a marker of poor health, but rather, can provide means of treating SCD prophylactically and on a more comprehensive level. One example is the use of penicillin prophylaxis in this vulnerable paediatric population.[40]

The strengths of this study include the robust total sample size, which allows us to measure the impact of SCD on a population that is representative of individuals across the USA. In addition, the NHIS contains data for children of multiple races, SES, SDoH, insurance coverage and different regions throughout the USA. This diversity and range of information makes it possible to compare children based on SCD status and substantiates the generalisability of our findings. In addition, we included available detailed information on the child's

Table 6 Health status and healthcare use for black children by sickle cell disease (SCD) status (N=21 565)

	Black children with SCD **n=(**170**)**		Black children without SCD **n=(**21 395**)**		AOR for SCD vs no SCD **(95% CI)**	**P value**
	Unweighted n	**Weighted %**	**Unweighted n**	**Weighted %**		
Health impact						
Limited in ability to crawl, walk, run or play	21	13.63	399	1.83	7.8 (4.2 to 14.4)	<0.001
Needed special equipment	3	1.45	263	1.23	1.3 (0.3 to 5.5)	0.707
Took prescription medication for at least 3 months	69	38.90	3139	14.00	3.9 (2.3 to 6.4)	<0.001
Reported health status fair or poor						
Better	58	36.73	5124	24.10	1.8 (1.2 to 2.9)	0.008
Worse	7	3.74	258	1.35	3.1 (1.0 to 9.2)	0.041
About the same	104	59.52	15 920	74.54	0.5 (0.3 to 0.8)	0.001
# of days school missed because of illness or injury						
0	24	26.78	5944	39.56	0.5 (0.2 to 0.9)	0.028
1–10	64	55.81	8749	57.10	0.9 (0.5 to 1.6)	0.758
11–20	13	9.38	407	2.42	4.1 (1.9 to 9.0)	<0.001
21–30	5	3.28	102	0.55	8.4 (2.9 to 24.3)	<0.001
31–40	0	–	17	0.01	–	–
40+	5	4.75	39	0.28	19.0 (5.5 to 65.5)	<0.001
Healthcare and special education services use (past 12 months)						
Saw a medical specialist	47	23.52	2507	11.56	2.5 (1.5 to 4.2)	0.001
Saw mental health professional	19	10.74	1417	7.38	1.1 (0.5 to 2.3)	0.900
Saw a therapist	16	12.56	1258	6.83	2.0 (0.9 to 4.1)	0.074
Saw an optometrist, ophthalmologist or eye doctor	50	32.18	4878	25.37	1.5 (0.9 to 2.5)	0.144
How many times have you seen a doctor (past 12 months)						
None	8	3.57	2022	9.25	0.2 (0.1 to 0.7)	0.009
1–5	111	69.22	16 583	79.38	0.6 (0.4 to 0.9)	0.019
6–9	22	13.30	1510	6.98	2.3 (1.2 to 4.5)	0.010
10–15	17	9.44	628	2.92	3.7 (1.9 to 7.3)	<0.001
16 or more	10	4.47	325	1.48	2.6 (1.0 to 6.8)	0.059
Multiple visits to emergency room (past 12 months)						
None	91	52.68	16 058	76.34	0.3 (0.2 to 0.6)	<0.001
1	30	17.42	3173	14.66	1.1 (0.6 to 1.9)	0.773
2–3	28	18.14	1572	7.15	2.8 (1.5 to 5.4)	0.002
4–5	11	7.31	266	1.15	5.1 (2.3 to 11.3)	<0.001
6–16+	10	4.45	148	0.69	7.3 (3.0 to 17.5)	<0.001
Had surgery or another surgical procedure	13	3.82	824	3.89	1.0 (0.4 to 2.0)	0.905
Healthcare barrier						

Continued

Table 6 Continued

	Black children with SCD **n=(170)**		Black children without SCD **n=(21 395)**		AOR for SCD vs no SCD **(95% CI)**	**P value**
	Unweighted n	**Weighted %**	**Unweighted n**	**Weighted %**		
Could not use telephone	5	4.42	361	1.79	1.7 (0.4 to 6.7)	0.431
Could not get an appointment	8	6.14	964	4.58	1.2 (0.5 to 3.0)	0.717
Waited too long at the doctor's office	4	1.33	808	3.87	0.3 (0.1 to 1.1)	0.064
No transportation	10	4.70	622	3.24	0.8 (0.3 to 2.1)	0.632
Could not afford prescription medicines	1	7.70	41	1.30	8.4 (2.4 to 29.4)	0.001

AOR, adjusted OR.

medical conditions, maternal education, health impacts, healthcare and special education services usage. Lastly, the survey weights provided by NHIS were used to weigh the sample to prevent skewing from categories with smaller sample sizes. Nonetheless, the results are also subject to possible limitations. The significant interactions observed between SCD status, SES and comorbid medical outcomes warrant further investigation and it is a call to action to design and implement sustainable evidence-based interventions that will address SCD management inequalities. Due to the small sample size of children with SCD, we could not examine the association between SCD status and healthcare access barriers for all children with SCD and black children with SCD compared with their counterparts. Further, although selection bias is not commonly seen in cross-sectional studies, it is possible that selection bias influenced some of the associations examined in this secondary analysis, mainly because we compared a small number of children with a condition that is not well represented in the general population of children across the USA. In addition, the data is collected through household surveys reported by parents/guardians, and the children's medical records are not accessible to confirm the information. Thus, the final survey results may be influenced by recall bias.[41] Previous NHIS studies of validity have shown that the child's parent/guardian likely underreports medical conditions related to chronic disabilities.[42] The NHIS data does not contain any information about SCD genotypes, which are associated with varying severities and frequencies of SCD and its complications. The survey contains a question about sickle cell anaemia (SCA) which may have been misinterpreted by parents/guardians who do not understand that SCD refers to a group of inherited blood disorders, while SCA refers to a specific type of SCD.[43] The prevalence of SCD was greatest in 2016, this could be due to better surveillance methods in preparation for approval of hydroxyurea for paediatric use, which occurred in late 2017. We excluded the 2006 data set from the analyses because the file was missing the person and income data sets needed to identify unique

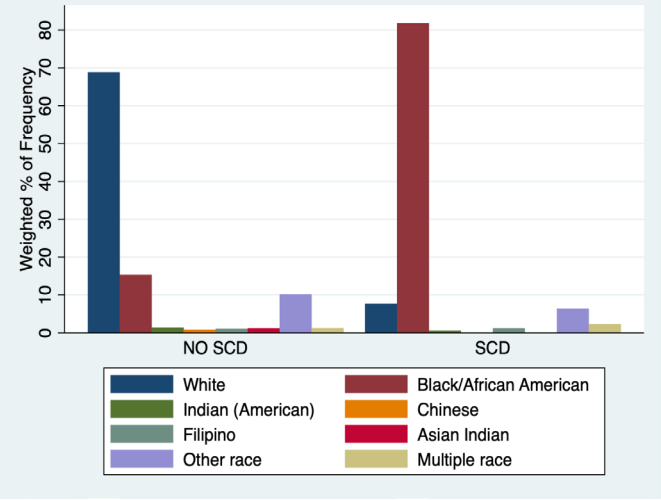

Figure 1 Sickle cell disease status by race/ethnicity data from National Health Interview Survey. SCD, sickle cell disease.

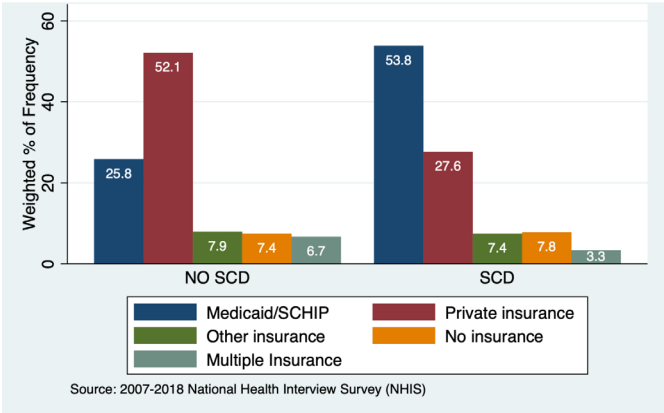

Figure 2 Insurance profile of respondents to the National Health Interview Survey based on sickle cell disease status. SCD, sickle cell disease; SCHIP, State Children's Health Insurance Program.

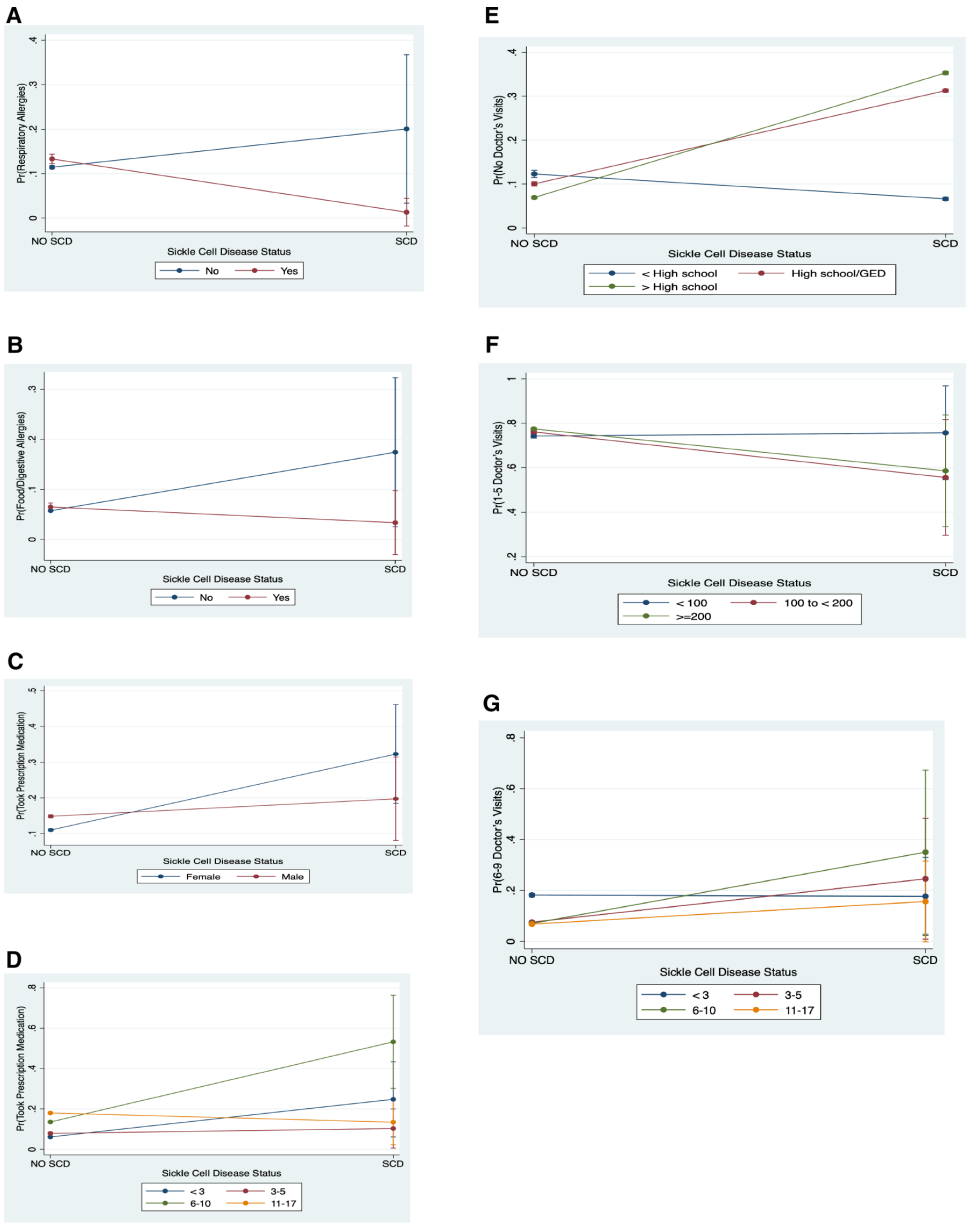

Figure 3 (A) Predictive margins of interaction between sickle cell disease status and other insurance (not Medicaid/private insurance) on **Respiratory Allergies** with 95% CI. (B) Predictive margins of interaction between sickle cell disease status and other insurance on **Food/Digestive Allergies** with 95% CI. (C) Predictive margins of interaction between sickle cell disease status and sex on **Took prescription medication for at least 3** months with 95% CI. (D) Predictive margins of interaction between sickle cell disease status and age on **Took prescription medication for at least 3** months with 95% CI. (E) Predictive margins of interaction between sickle cell disease status and mothers' education status on **no doctor's visits within 12 months** with 95% CI. (F) Predictive margins of interaction between sickle cell disease status and household income (% federal poverty level) on frequency of 1–5 doctor's visits within 12 months with 95% CI. (G) Predictive margins of interaction between sickle cell disease status and age on **6–9 doctor's visits within 12 months** with 95% CI. SCD, sickle cell disease.

children per household. Furthermore, a limited amount of SCD cases and other medical conditions were reported within the data set, affecting point estimates. However, the data proved useful when pooled to compare SCD and non-SCD children within the range of the corresponding CIs. A study by Wendler *et al*[44] assessing racial and ethnic minorities' willingness to participate in health research found that 'racial and ethnic minorities in the US are as willing as non-Hispanic whites to participate in health research'. Wendler *et al* found that the participation/uptake rate (defined as consent to participate in the study) for non-Hispanic whites, African Americans and Hispanics for NHIS was 83.6%, 81.1% and 81.7%, respectively. We recognise that there are challenges to recruiting communities of colour for research participation, especially for clinical research; and although the NHIS was a cross-sectional survey, still there may have been barriers to recruitment. A recent study by Webber-Ritchey and colleagues[45] provide great insight for recruitment strategies and lessons learnt when enrolling African American

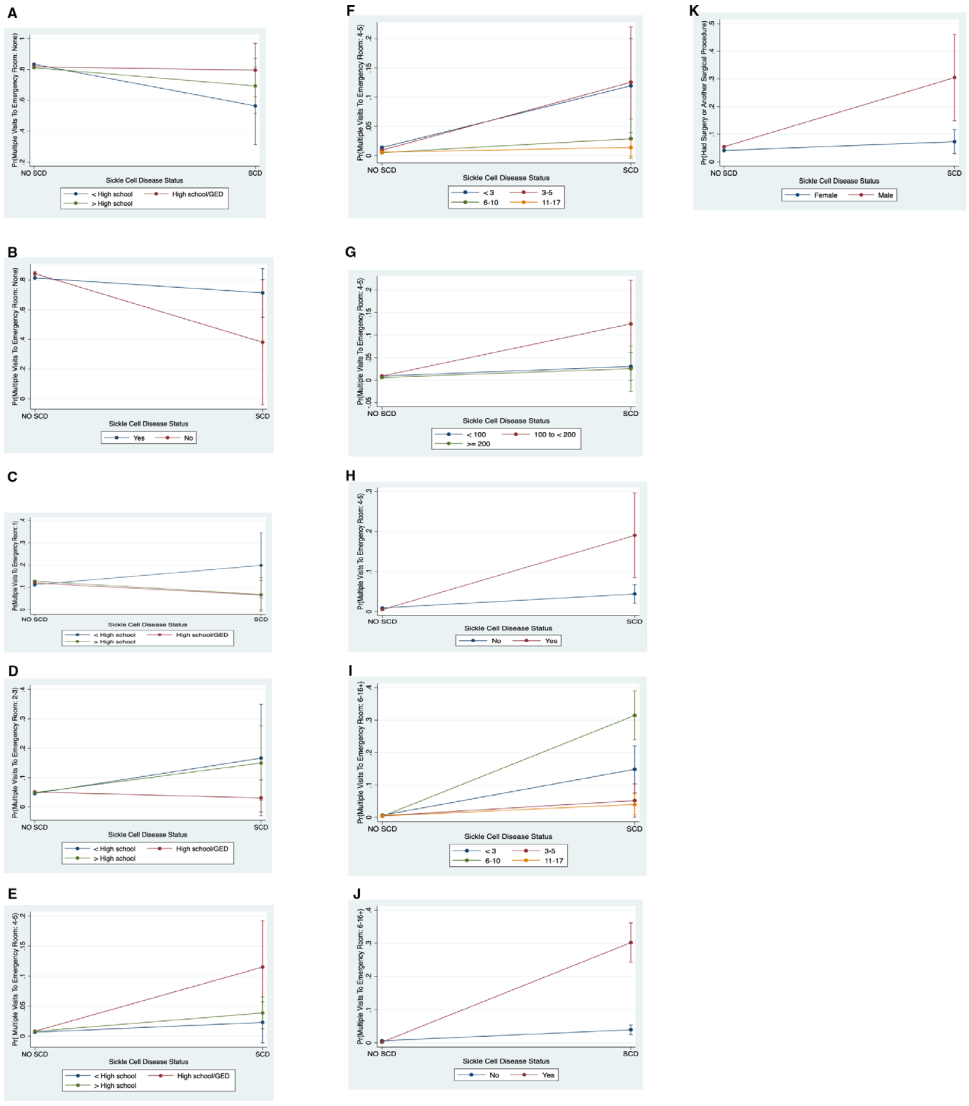

Figure 4 (A) Predictive margins of interaction between sickle cell disease status and maternal education on **Multiple visits to emergency room: none** with 95% CI. (B) Predictive margins of interaction between sickle cell disease status and insurance coverage on **Multiple visits to emergency room: none** with 95% CI. (C) Predictive margins of interaction between sickle cell disease status and maternal education on **Multiple visits to emergency room: 1** with 95% CI. (D) Predictive margins of interaction between sickle cell disease status and maternal education on **Multiple visits to emergency room: 2–3** with 95% CI. (E) Predictive margins of interaction between sickle cell disease status and maternal education on **Multiple visits to emergency room: 4–5** with 95% CI. (F) Predictive margins of interaction between sickle cell disease status and age on **Multiple visits to emergency room: 4–5** with 95% CI. (G) Predictive margins of interaction between sickle cell disease status and household income (% federal poverty level) on **Multiple visits to emergency room: 4–5** with 95% CI. (H) Predictive margins of interaction between sickle cell disease status and private insurance on **Multiple visits to emergency room: 4–5** with 95% CI. (I) Predictive margins of interaction between sickle cell disease status and age on **Multiple visits to emergency room: 6–16+** with 95% CI. (J) Predictive margins of interaction between sickle cell disease status and private insurance on **Multiple visits to emergency room: 6–16+** with 95% CI. (K) Predictive margins of interaction between sickle cell disease status and sex on **Had surgery or another surgical procedure** with 95% CI. SCD, sickle cell disease.

parents/caregivers of school-aged children in an online survey including using different recruitment strategies (flyers, emails, phone calls, in-person, etc) and providing online surveys to limit participant burden and provide flexibility for survey completion, when technology is not a challenge. NHIS data was collected via an in-person interview, which may have been burdensome to participants in terms of time and comfortability.

We found a low response rate to the healthcare access barrier questions. Future NHIS data collection should explore additional factors impacting healthcare access at the institutional, provider and patient level to better understand the context for SCD management in the USA, especially for blacks. There is clearly a need to strengthen the sickle cell disease national surveillance efforts in the USA.

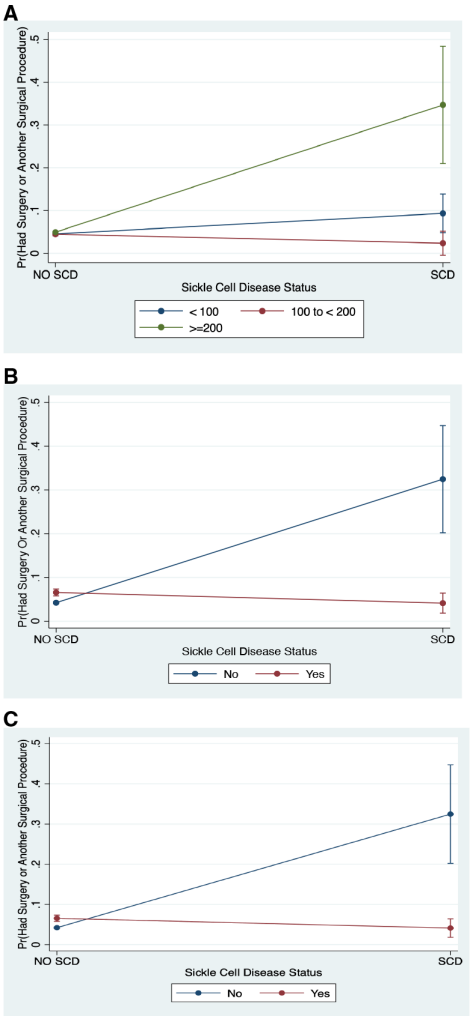

Figure 5 (A) Predictive margins of interaction between sickle cell disease status and household income (% federal poverty level) on **Had surgery or another surgical procedure** with 95% CI. (B) Predictive margins of interaction between sickle cell disease status and public insurance on **Had surgery or another surgical procedure** with 95% CI. (C) Predictive margins of interaction between sickle cell disease status and private insurance on Had surgery or another surgical procedure with 95% CI. SCD, sickle cell disease.

CONCLUSION

Our study findings indicate that children with SCD experience significant comorbid conditions. Moreover, black children with SCD have more SCD-related comorbidity, more ER visits and worse health status than black children without SCD. This creates the urgency to address the residual information gaps in health burden for black children with SCD, by identifying 'beneficial' healthcare services (non-emergency services) to improve access and quality of life. Future research should focus on strategies to scale evidence-based interventions for underserved populations with SCD. The study results suggest that this group could significantly benefit from investments in interventions to improve health outcomes and also address SDoH. Moreover, implications for paediatric clinical practice are significant and findings may urge clinicians to make concerted efforts to reduce healthcare disparities.

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Patient consent for publication Not applicable.

Ethics approval Not applicable.

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