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A Message from the President

COVID-19 remains an evolving crisis and is Killing Black People Unequally. The current state of public health with the novel coronavirus demands that at-risk populations need to be taken care of. This pandemic has amplified unequal treatment not only in access to healthcare for racial and ethnic minority communities but in every thread of our livelihood. We are quite familiar with healthcare disparities and the numerous programs, research, and policies that have evolved as a result of the 2003 report Unequal Treatment: Confronting Racial and Ethnic Disparities by the Institute of Medicine, which found that race and ethnicity remain significant predictors of the quality of healthcare a person receives. COVID-19 has revealed the paucity in the nationwide improvement of these initiatives. The Black Community is particularly suffering and accounts for a disproportionate number of the COVID-19 cases and fatalities in the United States. The disparities are striking but not surprising. Access to care remains insufficient in Black Communities. Many underlying diseases in the Black Community have been targeted as leading to worse outcomes. These diseases are often expressed as ones that can be solely modified by the individual. More exercise, less high caloric diets, etc. But we know individuals and community deficits are a combination of systemic, operational, judicial, and social inequalities that we must face. Biased systems that work in harmony can no longer be blind to a newborn hereditary disease such as sickle cell disease because it predominately impacts a race that continues to experience abuse, harm, and neglect. Healthcare is not a trusted and sacred place for individuals with sickle cell disease. Those affected continue to be treated as less than.

Early reporting of clinical data of individuals with sickle cell disease infected by the novel coronavirus shows that individuals with sickle cell disease are at increased risk for mortality and poor health outcomes. The dearth of access to care is widespread and exposed in ways that will no longer be muted, whispered about, or disguised.

A comprehensive, multi-race, multi-ethnic strategy is needed to eliminate these inequities.

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The Editor-in-Chief of Journal of Sickle Cell Disease and Hemoglobinopathies (JSCDH) would like to thank all reviewers for sharing their time, knowledge and expertise with JSCDH's authors in the evaluation of their work, without which it would be impossible to maintain the standards of our Journal.

TOP ABSTRACTS

Presenting: Friday, March 5, 2021 at 11:30am

JSCDH-D-21-00001

Food Deserts are Associated with Acute Care Utilization in Preschool Children with SCD

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Background: In the US, individuals with sickle cell disease are frequently poor and experience multiple acute and chronic complications (vaso-occlusive crisis) (Wang W. Lancet 2011) due to high social and nutritional vulnerabilities (McCaskill M. Nutrients 2018, Martyres D. PBC 2016, Schall J. J Pediatr 2004). While, infants and young children (age < 6) with SCD are particularly vulnerable to malnutrition that contributes to poor health outcomes (e.g., developmental delays, cognitive impairment), the environmental components leading to decreased food access have not been investigated relative to the impact on their healthcare outcomes. We tested the hypothesis that restricted access to healthy food sources is associated with increased SCD-related acute care utilization among children younger than age 6 years, despite treatment with hydroxyurea.

Methods: Participants were recruited from the IRB-approved longitudinal clinical cohort study, Sickle Cell Clinical Research and Intervention Program (Hankins J. PBC 2018). Home addresses were mapped to census-tract environmental data from the US Food Access Research Atlas (USDA ERS 2017). Food deserts were defined as "low income census tracts where at least 33% (minimum of 500 people/tract) of the population live >1.0 (urban area) or >10 (rural area) miles from a grocery store or a supermarket" (Food Access, USDA ERS 2019). Three main outcomes: emergency department (ED) visits, hospitalizations, and acute care utilization (ACU=ED + hospitalizations) from a vaso-occlusive event, were collected from birth to age 6 and analyzed as cross-sectional outcomes at age 6-years. Generalized linear models (GLM) were used to associate environmental factors as continuous and categorical variables with the outcomes adjusted for sickle genotype and hydroxyurea exposure.

Results: 523 children with SCD, all African American, were included. The median age at last follow-up was 5.5 years (range 1– 6), 51.7% were girls (Table 1). A total of 33.5 % of the studied population resided in census tracts considered food deserts. The average distance to the nearest supermarket from participants' household was 2.8 miles (Table 1). Participant neighborhoods had on average 14.7% unemployment rate, while 30.8% of individuals were under the federal poverty threshold and received Food and Nutrition Services. Among the tracts where the population was considered low income, 9% did not own a car. Living in a household without a vehicle and located >0.5 miles from a supermarket was associated with increased hospitalizations and acute care utilization (ACU) (Figure 1). The odds ratio (OR) of experiencing >0 hospitalizations or ACU were 1.3 (95%CI: 1.0-1.8) or 1.5 (95%CI: 1.1-2.0), for those living in a household without a vehicle and >0.5 miles from a supermarket, respectively. Living in a household with children and >1.0 mile from a supermarket was associated with high risk of experiencing >0 hospitalizations (OR: 1.5; 95%CI: 1.2-1.8) and >0 ACU (OR: 1.3; 95%CI: 1.1-1.7) (Figure 2). The accuracy of predicting a SCD-related acute event by age 6 years significantly improved when adding markers of poor food access to the predictive model (AUC increase: ≥0.06, p=0.01) (Figure 3).

Conclusions: Living in food deserts limits access to affordable and nutritious foods. Food deserts are associated with poor health outcomes among pre-school children with SCD. The prediction of acute care utilization in young childhood increases when food access is considered. Treatment with hydroxyurea did not mitigate the effects of reduced food access on the frequency of acute care utilization of young children with SCD.

Table 1. Participants and their Neighborhood Characteristics

		Sickle Genotype			
		All (N=523)	HbSC/Sβ⁺thal/ Other (N=197)	HbSS/Sβ⁰thal (N=326)	p-value
Patient/Parent-Level Characteristics					
Demographics and Healthcare Utilization	Sex (Female)	270 (51.7)	105 (53.3)	165 (50.6)	0.59
	Mean Age (SD) Years	5.5 (1.3)	5.53 (1.2)	5.48 (1.3)	0.96
	Treatment with Hydroxyurea (%)	144 (27.5)	11 (5.6)	133 (40.8)	**<0.0001***
	Mean duration of Hydroxyurea therapy (SD) years	0.67 (1.33)	0.1 (0.6)	1.02 (1.6)	**<0.0001***
	Mean number of all acute care visits (ED + hospitalizations) per year (SD)	0.33 (0.57)	0.21 (0.5)	0.4 (0.6)	**<0.0001***
	Mean number of ED visits per year (SD)	0.17 (0.35)	0.13 (0.3)	0.19 (0.4)	**0.025**
	Mean number of Hospitalizations per year (SD)	0.16 (0.33)	0.08 (0.2)	0.21 (0.4)	**<0.0001***
	Study population mapped to a census tract (N)	460 (87.9)	184 (93.4)	276 (84.7)	0.28
Community Level Characteristics (Per Census Tract)					
General Characteristics	Mean % of population below poverty rate (SD)	29.05 (14.1)	28.86 (14.9)	29.18 (13.5)	0.82
	Mean % of unemployed population (SD)	14.66 (7)	14.91 (7.3)	14.49 (6.8)	0.49
	Mean Household Income in 2015 US dollars (SD)	37375.31 (19184.9)	38700.74 (21264.3)	36445.71 (17571.5)	0.85
	Mean % of children (SD)	28 (5)	28 (5)	29 (5)	**0.00083***
Educational Attainment	Mean % individuals with high school degree (SD)	28.46 (7.3)	28.5 (7.9)	28.43 (6.8)	0.59
	Mean % of individuals with bachelor's degree (SD)	11.79 (7.9)	12.7 (8.7)	11.16 (7.3)	0.13
	% of individuals with bachelor's degree, n (%)				0.5
	< 10%,	181(48.3)	70(45.4)	111(50.2)	
	≥ 10%, <20%	147(39.2)	60(39.0)	87(39.4)	
	≥ 20%, <30%	24(6.4)	12(7.8)	12(5.4)	
	≥ 30%,	23(6.1)	12(7.8)	11(5.0)	
Transportation and Food Access	Mean % of individuals on Supplemental Nutritional Assistance Program (SNAP) (SD)	30.78 (14.2)	30 (15.2)	31.33 (13.4)	0.43
	Mean distance to a supermarket (miles) (SD)	2.82(1.5)	2.83(1.5)	2.81(1.5)	0.96
	Mean % of census tract population living >0.5 miles from supermarket (SD)	74 (25)	74 (25)	75 (25)	0.62
	Mean % of census tract population living > 1.0 miles from supermarket (SD)	34(36)	33(35)	35(36)	0.71
	Mean % of households without vehicle (SD)	12(10)	12(10)	13(9)	0.14
	Mean % of households without a vehicle living > 1.0 miles from supermarket (SD)	3(6)	3(5)	4(6)	0.17
	Mean % of low-income households living >0.5 miles from supermarket (SD)	37 (21)	36(21)	38(20)	0.14
	Mean % of households with children living >1 mile from supermarket (SD)	10 (10)	9 (10)	10 (11)	0.5
	Mean transit access index (0 to 100) (SD)	20 (14.4)	20.8 (16.1)	19.44 (13.1)	0.84
	Mean % of households without a vehicle living >0.5 miles from supermarket (SD)	9 (8)	8 (9)	9 (8)	0.064
	% of households without a vehicle living >0.5 miles from supermarket, n (%)				0.12
	< 10%,	292(63.5)	127(69.0)	165(59.8)	
	≥ 10%, <20%	133(28.9)	42(22.8)	91(33.0)	
	≥ 20%, <30%	22(4.8)	9(4.9)	13(4.7)	
	≥ 30%,	13(2.8)	6(3.3)	7(2.5)	
	Mean % of low-income households living > 1.0 miles from supermarket (SD)	16(20)	14(19)	17(21)	0.32
	% of low-income households living > 1.0 miles from supermarket, n (%)				0.81
	< 10%,	266(57.8)	109(59.2)	157(56.9)	
	≥ 10%, <20%	96(20.9)	39(21.2)	57(20.6)	
	≥ 20%, <30%	75(16.3)	29(15.8)	46(16.7)	
	≥ 30%,	23(5.0)	7(3.8)	16(5.8)	
Health and Safety	Mean distance to nearest police station (miles) (SD)	1.95 (1.0)	2.03 (1.1)	1.88 (0.9)	0.2
	Mean distance to nearest fire station (miles) (SD)	0.91 (0.5)	0.93 (0.5)	0.9 (0.42)	0.79
	Mean distance to nearest hospital (miles) (SD)	2.86 (1.4)	3.01 (1.4)	2.76 (1.4)	0.073
	Mean census block Blight rating (1 to 5) (SD)	1.92 (0.5)	1.84 (0.4)	1.97 (0.5)	0.11

Notes: SD = standard deviation, ED = emergency department. * False discovery rate (FDR) adjusted p-values < 0.05.

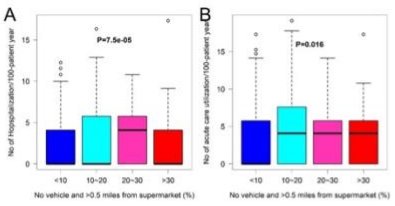

Figure 1. Association between distance to a supermarket without vehicle ownership and the number of acute care utilizations by age 6. The x-axis is the % of household in the census tract that did not have a vehicle and was located beyond 0.5 miles from a supermarket divided based on three cutoff points of 10%, 20% and 30%. Y-axis is the square root of the number of hospitalizations (A) and acute care utilizations (B) per-100-patients per year. There is a significant increase in hospitalizations (A) and all acute care utilization (B) relative to the number of households in the census tract without a vehicle and >0.5 miles from a supermarket, such that the greater the number of household with decreased food access, the more acute events. P-values were calculated from generalized linear model with quasi-Poisson link function with adjustment for hydroxyurea exposure and SCD genotype by analyzing the categorized environmental variable as a dose effect of 0, 1, 2 and 3. False Discovery Rate (FDR) adjusted p-values for A is <0.05 and for B is =0.1.

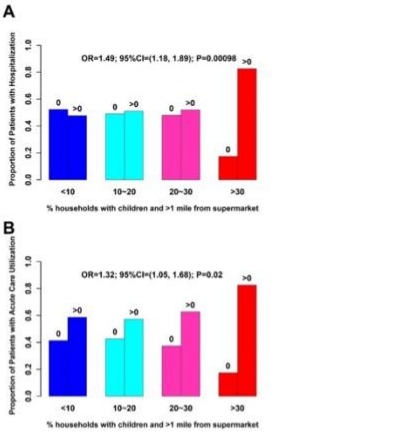

Figure 2. Association between households with children who live beyond 1 mile from a supermarket and acute care utilization occurrence by age 6 years. The x-axis is the % of households in the census tract that had children and was located >1.0 miles from a supermarket, divided based on 10%, 20% and 30% of households in the census tract with these attributions. The Y-axis is the number of patients with hospitalization (A) or acute care utilization (B) by age 6 years, with zero denoting no events and >0 denoting one or more events by age 6. P-values, odds ratio (OR) and 95% confidence intervals (CI) were calculated from generalized linear model with binomial link function, adjusting for hydroxyurea exposure and sickle genotype by analyzing the categorized environmental variable as a dose-effect of 0, 1, 2 and 3. FDR adjusted p-values for A and B are <0.07.

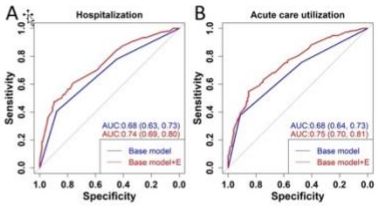

Figure 3. Receiver Operator Characteristic Curves of the Prediction of Acute SCD-Related Events by Age 6 using environmental markers of poor food access. The base model includes covariates of SCD genotype and hydroxyurea exposure. E (environmental) variables include % living in a household with children who were located beyond 1.0 mile from a supermarket and % of individuals with bachelor's degree (panels A and B), or living in a household that did not have a vehicle and located beyond 0.5 miles from a supermarket (panel B). Environmental variables were analyzed as groups of four categorical variables based on incremental proportions of households or individuals with the environmental attributes.

JSCDH-D-21-00002

Prevalence of Neuro-Developmental Deficits in Young Children with Sickle Cell Disease

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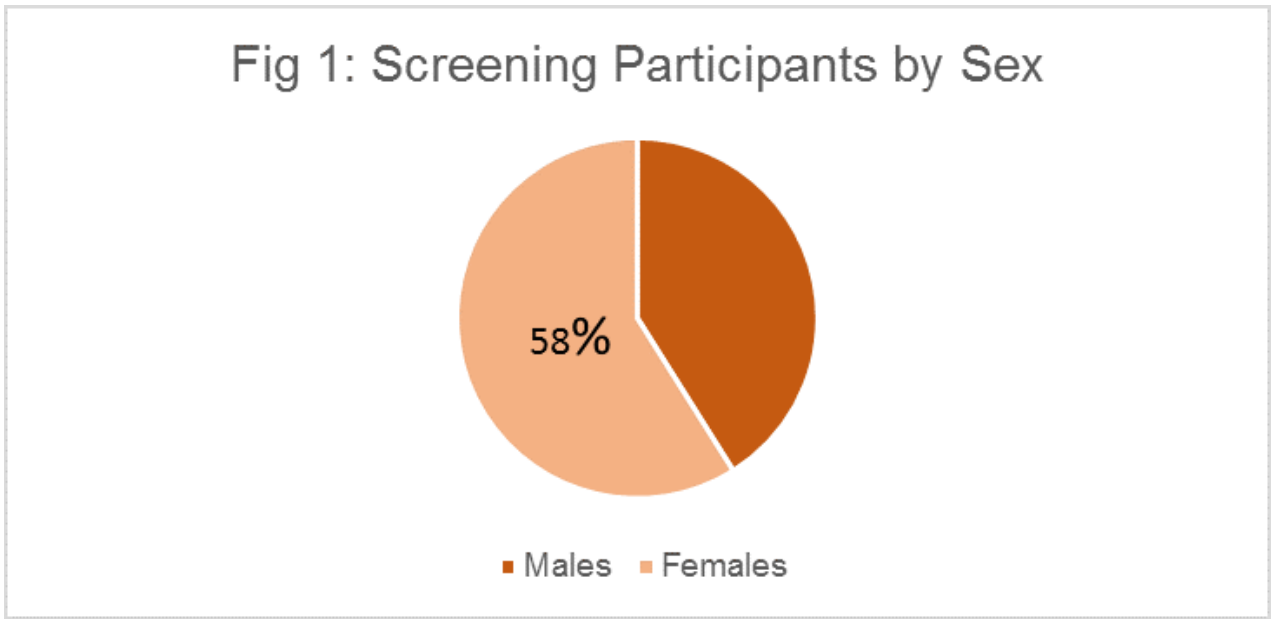
Background: Sickle cell disease (SCD) is an inherited disorder of red blood cells that leads to poor oxygen supply and multi-organ damage. It affects nearly 100,000 Americans, majority of whom are of Black or African American ancestry (CDC, 2020). Neurologic complications include cerebral vasculopathy, silent infarcts and overt strokes which can disrupt normal neurologic development in childhood, and lead to cognitive deficits and disability in adulthood. In addition, young children with sickle cell disease are often dealing with the additional burden of healthcare disparities, socio-economic challenges and inadequate primary care. As a result, many neuro-developmental delays are under-recognized or missed in early childhood. The Parents' Evaluation of Developmental Status, or PEDS group of tests, are evidence-based screening tools for children aged 0 –8 years that identify parental concerns regarding language, motor, self-help and academic skills as well as socio-emotional and mental health status. These tools can be used to identify developmental delays in children with SCD who may have inconsistent primary care follow up. We sought to describe the neuro-developmental deficits identified in our patient population using these tests

Methods: The PEDS Developmental Milestone (PEDS: DM) screening test was performed by the social worker during her evaluation at comprehensive sickle cell clinic visits. Screenings were administered routinely to all children with SCD under 60 months (5 years) old, and to children with parent-identified concerns between 5 and 8 years old. Testing took place in person and on campus at the Children's Hospital of Georgia in Augusta, as well as via telephone for children who were seen in the satellite clinics across rural South Georgia (Waycross, Albany, Valdosta, Athens and Dublin). Age-appropriate developmental screening was performed for quarterly milestones in infants (3, 6, 9 and 12 months) and yearly milestones in children over the age of 12 months. Optional Modified Checklist for Autism in Toddlers, or M-CHAT, screening was available for children with parent-identified concerns.

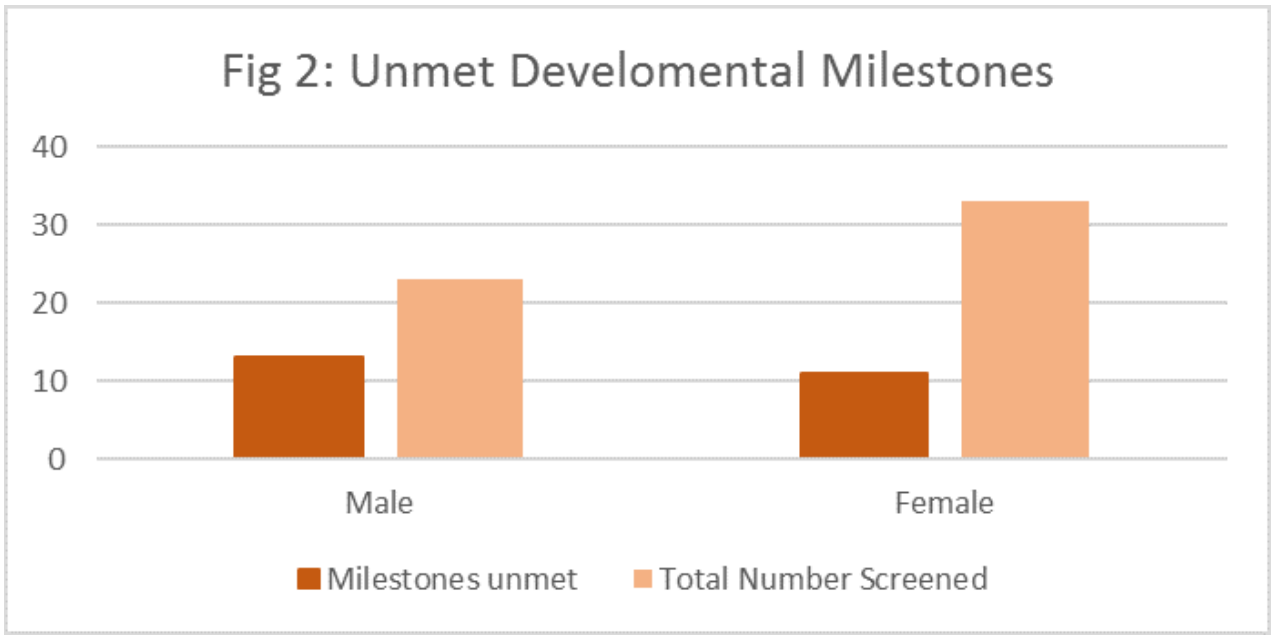
Results: A total of 62 developmental screening tests were performed on 56 children, with 6 children having more than one test. Age range of children who underwent screening was 3 – 50 months (median: 23 months). Thirty-three out of 56 children (58.9%) were females and 23 were males (Fig 1). Fifty-four children identified as African American (96.4%), and the other 2 identified as Hispanic. On review of screening results, 24 out of 56 (42.8%) children had unmet milestones in at least one domain, with 12 children (21.4%) having unmet milestones in 2 or more domains. A sex predilection was noted with 13 out of the 23 male children (56.5%) screened having unmet milestones when compared to 11 of 33 female children (33.3%) (Fig 2).From the population experiencing unmet milestones, the most common deficits were in fine (45.8%) and gross (33%) motor delays. In addition, children experienced delays in expressive language (25%), self-help (20.8%) and social-emotional skills (16.7%) (Fig 3).

Conclusion: Our developmental screening identified a high prevalence of neuro-developmental delays and unmet milestones in young children with SCD, about 43% in our population, with fine and gross motor delays being the most commonly identified deficits. Apart from underlying SCD, male children appeared to be at higher risk than female children for developmental delays. The reasons for this are unknown and beyond the scope of the screening tool. Although our data is limited by small sample size, it highlights the importance of routine developmental screening to identify and manage early neurologic deficits especially in young children with SCD. Future studies should be directed towards assessing factors, if any, that may contribute to higher risk in young boys, and directing appropriate resources to this at-risk population.

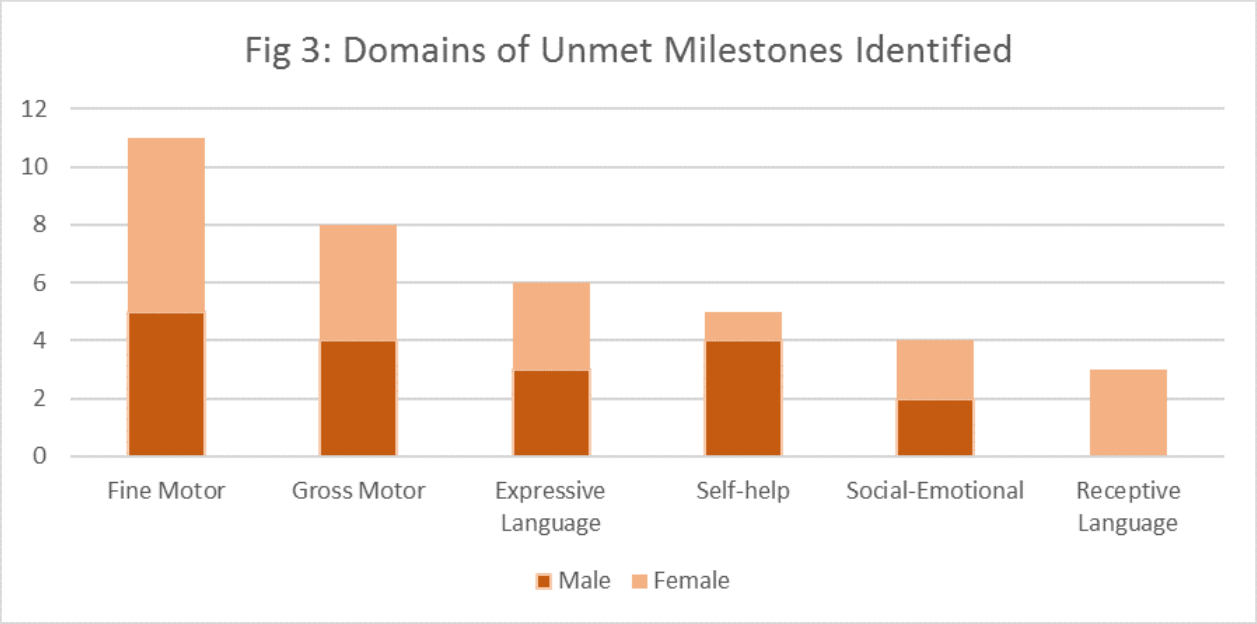
Sex Distribution of Screening Participants



Unmet Developmental Milestones in SCD Population by Sex



Domains of Unmet Developmental Milestones



JSCDH-D-21-00003

RELATIVE PREVALENCES OF BEHAVIORAL DIAGNOSES IN HIGHEST-UTILIZING SCD PATIENTS

Authors: Wally R. Smith, MD, Benjamin Jaworowski, BS, Shirley Johnson, BA, LSW, Thokozeni, Lipato, MD, Daniel M. Sop, BS, MS

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Background: Compared to other adults, those with anxiety, depression, alcoholism, schizophrenia, or substance misuse are known to utilize more health care. These diagnoses are prevalent in adults with sickle cell disease (SCD), who are overrepresented among high utilizers of emergency departments and inpatient beds. Behavioral interventions targeting health care utilization may wish to focus on the highest impact behavioral diagnoses first. We therefore studied patients in our Adult SCD Medical Home to determine which behavioral diagnoses had the highest relative prevalence among highest utilizers vs among all patients.

Methods: Virginia Commonwealth University Health system claims data from 2018-2019 were analyzed for the presence of ICD codes for SCD (n=746), along with one of the following: alcoholism, cannabis related disorders, sedative/hypnotic/anxiolytic disorders, stimulant related disorders, schizophrenia, mood disorders, anxiety, dissociative, and stress-related disorders, and personality and behavioral disorders. The top 50 highest utilizing SCD adults were determined by ranking in descending order in the preceding year: total charges, followed by inpatient discharges, then inpatient returns within 30 days, then ED use, then ED returns within 3 days. We compared the highest utilizers with all patients for the prevalence of each reported behavioral diagnosis. We then calculated a prevalence ratio for each diagnosis.

Results: The disorders which were most disproportionately prevalent in highest utilizers, in order of rank from first, to last, are shown in the Table. Anxiety and mood disorders were by far the most prevalent behavioral health disorders. However, sedative disorders, cannabis disorders, and schizophrenia and psychoses were the most disproportionately represented disorders among highest-utilizing patients. Cocaine, alcohol, personality, and other stimulant disorders were rare and not disproportionately represented among high utilizers.

Conclusions: In this sample of SCD adult patients, the higher impact on utilization of substance use vs. depression and anxiety, despite a lower prevalence, may be somewhat surprising. We hypothesize patients might be chemically coping with pain and underlying behavioral disorders by using sedative/anxiolytics and cannabis. Programs aimed at reducing utilization in SCD adults might be better served by first focusing on treating non-opioid substance misuse and psychoses, although mood disorders and anxiety should be managed.

	Prevalence		
Disorder	*Highest Utilizers, % (n=50)*	*All, % (n= 746)*	*Prevalence Ratio, Highest Utilizers/All*
Sedative/hypnotic/anxiolytic	4	0.8	5.00
Cannabis	22.7	6	3.78
Schizophrenia, psychosis	5.3	1.5	3.53
Mood	44	17.8	2.47
Anxiety	41.3	18.6	2.22
Cocaine	5.3	2.8	1.89
Alcohol	2.7	1.5	1.80
Personality	2.7	1.9	1.42
Other Stimulant	0	0.3	0.00

POSTERS

JSCDH-D-21-00004

BIDIRECTIONAL RELATIONSHIP BETWEEN SICKLE CELL DISEASE AND FOOD INSECURITY: SCOPING REVIEW

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Background: A growing body of research has shown that social determinants of health, such as food insecurity, can worsen health outcomes of those with chronic illnesses. According to the USDA, food insecurity is defined as "a household-level economic and social condition of limited or uncertain access to adequate food" affecting approximately 15.8 million (12.7%) of US households. In the United States, sickle cell disease (SCD)--the homozygous inheritance of a point mutation within the beta-globin chain of hemoglobin-- affects between 80,000-100,000 people. Adequate nutrition can influence the pathophysiology of SCD; individuals with SCD who are undernourished are more likely to have impaired immune function, poor ulcer healing, and disease exacerbation. Currently, limited research has explored the relationship between food insecurity and health outcomes in those with SCD, highlighting the importance of conducting research on food insecurity among those with SCD. The objectives of the current study are to understand the 1) prevalence of food insecurity among individuals with SCD; 2) impact of psychosocial factors on food insecurity among individuals with SCD; and 3) effect of food insecurity on SCD disease severity and impact of SCD severity on food insecurity3. Through a scoping review, we examined evidence from various food insecurity and SCD studies elucidating the bidirectional relationship between SCD and food insecurity.

Methods: For this review, we conducted a comprehensive search for peer reviewed manuscripts using the following databases: PUBMED, Embase, Cochrane, CINAHL, Scopus, and Web of Science, through November 2020. The search included the terms "food security", "food insecurity", "sickle cell" and "sickle cell disease". All identified manuscripts were screened for inclusion.

Results: From the database searches, thirty-five articles were identified. Thirty of these articles were not reviewed as they did not measure food security or food insecurity. Five articles were then reviewed for descriptive analysis. Of the five published studies, the publication dates ranged from 2018 to 2020 and included populations from the US and Brazil. These studies included one exploratory study, three cross-sectional studies, and one retrospective cohort study. Analysis of selected articles revealed several key findings. The prevalence of food insecurity among individuals with SCD were 21.3% , 35%, 62%, 40.6 %, and 62.2 % . A significant difference was established between caregiver and child assessment of food insecurity. Household food insecurity for families with a child with SCD was 21.3% while 45.8 % of surveyed children with SCD were food insecure. Interestingly, Gruntoad et al. found that 35% of their participants with SCD had some level of food insecurity but reported average quality of life (QOL) score. An inverse relationship was found between food insecurity and social support, particularly in families of children with severe SCD. There was a significant difference in emergency department reliance (EDr) and EDr increased by an average of 7.7 points per additional social determinants of health (SDOH) which included food insecurity.

Conclusion: The limited findings from this scoping review reveal a relationship between SCD and food insecurity in both adult and pediatric populations. Social support is highlighted as a protective factor against food insecurity. The severity of SCD may increase EDr and cost of hospitalization, influencing food insecurity.However, there is a critical lack of research among food insecurity in SCD communities, particularly in how both may exacerbate each other bidirectionally due to the financial burden of chronic illness and resulting stress. Additional research using rigorous methodologies is required to better understand the bidirectional relationship between SCD and food insecurity and to develop policies to alleviate food insecurity among individuals with SCD and their families.

JSCDH-D-21-00005

COVID-19 Symptoms vs Risk Perception and Care Delay in Sickle Cell Disease

Authors: Wally R. Smith, MD, Benjamin Jaworowski, BS, Shirley Johnson, BA, LSW, Thokozeni, Lipato, MD, Daniel M. Sop, BS, MS

Affiliation: *Virginia Commonwealth University*

Background: Sickle cell disease (SCD) patients were federally declared as more vulnerable to COVID-19 early in the pandemic. SCD patients often delay or avoid needed care, though they see themselves as vulnerable. We therefore implemented a telephone survey to assess reported COVID symptoms in our Adult Sickle Cell Medical Home, as well an email survey to assess attitudes about COVID-related risks as well as care delay or avoidance behavior.

Methods: Fifteen staff in the at Virginia Commonwealth University conducted a quality improvement (QI) telephone survey, with IRB waiver, of patients age >17 with SCD (N=622) from June 1-June 19, 2020. We asked the following yes/no screening questions about possible COVID-19 infection in the prior 14 days: fever>100 F, cough, difficulty breathing, unexplained shortness of breath, sore throat, unexplained muscle soreness, COVID-19 testing, COVID positivity, contact with anyone who tested positive for COVID-19. Telephone respondents were then invited to complete an email survey of 19-33 items, depending on their response pattern. Reported here are survey items we drew from the National Health Interview Survey, the Adult Sickle Cell Quality of Life Measurement Quality of Care survey and new items drafted to tap COVID risk perception. We also used hospital claims data to catalog ED use of these 622 patients from September 2019 to August 2020.

Results: For QI purposes, we set a threshold of >2 COVID-associated symptoms, or the presence of fever, as requiring further testing or monitoring immediately. Of 354/622 adults (153 M, 201 F) approached by phone who responded, only 3/354 (1%) met threshold, although 6/353 (2%) had had testing for COVID-19. Of 219 patients who had valid email addresses, 64 (29%, 21 M, 43 F) returned email surveys in time. Results are shown in Table 1. Over a third of SCD adults were already frequently managing VOCs at home prior to COVID. In the prior 14 days about half of patients had had a VOC at home, but very few visited the ED for their VOC. A quarter avoided “needed care”; just over half delayed “needed care” for at least a day. Table 2 confirms a sharp decline in emergency utilization in the sample starting in March 2020.

Conclusion: In SCD adults, pre-COVID care delay behavior continued or perhaps worsened early in the COVID pandemic. Despite amazingly low COVID symptom or infection rates, self-perceived risk of COVID susceptibility or morbidity was high. Future studies could compare delays of care and perceptions of COVID risk in SCD adults versus non-vulnerable adults, and could compare these behaviors and attitudes in SCD adults pre- vs. post-vaccine availability.

Frequency Response Items*	%
In the past 12 months:	
Managed "pain attack" at home ≥4 times	35.9
In the past 14 days:	
Crisis for at least one day	46.9
Visited ED for pain	6.3
Avoided "needed care"	25.8
Delayed "needed care" by at least a day	59.0
Advised by clinician to come for medical care but refused or did not come	6.3
Scale Response Items	**Weighted mean (0-10 scale)**
Worry, COVID infection overall	6.25
Worry, COVID infection because of going for medical care	3.9
Worry, more at risk of COVID because of SCD	6.31
Worry, will fare worse than others if COVID infected	6.97
Taking more medication to calm down because of COVID	1.6
** n=64 except Forwent "needed care," n=62; Delayed "needed care", n=61*	

Mon-Yr*	ED Use, No. pts.	ED Use per 1000 pts
Sep-19	161	290.6
Oct-19	159	287.0
Nov-19	157	283.4
Dec-19	145	261.7
Jan-20	130	234.7
Feb-20	159	287.0
Mar-20	105	189.5
Apr-20	68	122.7
May-20	84	151.6
Jun-20	88	158.8
Jul-20	97	175.1
Aug-20	89	160.6
**Virginia COVID lockdown on 3/23/20; ED= Emergency Department.*		

JSCDH-D-21-00006

CREATING AN EDUCATIONAL MODULE TO IMPROVE PROVIDER ATTITUDES TOWARDS SICKLE CELL PATIENTS

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Background: Sickle cell disease (SCD) exemplifies systemic racism in health care. For many SCD patients, the worst inequity is the staff's negative attitude when seeking pain care. Too often, they hear the words "sickler," "drug seeker," or "frequent flyer." These depersonalizing and derogatory terms reflect a negative provider attitude. For many, coming to the ED for treatment is a balance between their dignity or their life. Negative provider attitudes directly impact outcomes by delaying care and increasing hospitalizations and complications, and cost. We all want to do what is right. Sometimes our attitudes toward patients are affected by our implicit bias, negative experiences with patients' behavior, and even the institute's culture -the hidden curriculum. Stories from patients and providers affect provider attitudes dramatically. Though successful in pilot programs, they are not a standard part of the curriculum. Training by learning the stories likely will change the provider's attitude.

Methods: Through our pediatric resident quality improvement learning collaborative, we create an educational module based on the compelling video – "They don't believe me" and analyze a clinical note containing stigmatizing words. The model will be evaluated and modified through focus groups of residents and a patient advisor. We will use both quantitative and qualitative measures to determine the effectiveness of our module. Specifically, we will use: 1. The Positive Provider Attitudes toward SCD Patients Scale (PASS), a validated tool to evaluate the educational module's pre-and post-effects on resident attitude. 2. Focus groups and semi-structured interviews to determine staff perception of effectiveness. 3. Analysis of data from "PeopleSoft," including the numbers completing the module and changes in attitude. 4. Surveys of patients' confidence that they will be treated with dignity and respect in their next encounter for pain, performed in the follow-up clinic.

Results: This project is a work in progress. At our institution, 45 pediatric, 63 emergency medicine, and 120 internal medicine residents are participating. This education module is being implemented through "PeopleSoft," an educational platform used across our 11 hospitals. We expect it to reach over 1000 residents within our public hospital system. If successful, this model can be easily replicated and sustained as part of any e-learning system.

Conclusion: We are creating an educational module to improve provider attitudes to SCD patients for use across residency specialties and institutions through the "PeopleSoft" platform. Ultimately, we expect this intervention will lead to patient confidence that they will always be treated with dignity and respect.

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FAMILY-CENTERED COMMUNICATION IN PEDIATRIC SICKLE CELL DISEASE

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Background: Individuals with sickle cell disease (SCD) experience disparities in access to high quality, comprehensive care, which contribute to poor health outcomes. Research suggests that patient/family-provider relationships are an important indicator of healthcare quality and can influence disease self-management. Specifically, the Patient Centered Communication (PCC) framework (Epstein & Street, 2007) holds that patient/family-centered communication (e.g., eliciting, understanding, and validating patients' and families' perspectives within their unique psychosocial contexts) contributes to improved self-efficacy for disease management, adherence, and health outcomes. While the PCC framework has demonstrated utility in other pediatric populations (e.g., cancer, Type 1 diabetes), it has not yet been applied in the context of pediatric SCD. This study aimed to use this framework to examine patient and family perceptions of communication with pediatric SCD healthcare providers.

Methods: 17 caregivers (82% mothers, 94% Black/African American) and 8 patients aged 13-19 years (M=16.50, 62% female) completed individual semi-structured interviews as part of a larger study examining treatment decision making. The PCC framework informed a preliminary codebook, and both inductive and deductive analyses are ongoing. Two raters are independently coding interviews; discrepancies will be resolved via group discussion to consensus. Thematic content analysis will summarize family perspectives regarding communication with healthcare providers.

Results: Preliminary findings suggest that for youth with SCD and their caregivers, family-centered communication reduces patient/family distress, supports disease self-management efforts, facilitates information exchange and decision making regarding treatment, and fosters positive and trusting relationships with providers.

Conclusion: The PCC framework is a useful model for understanding patient/family-provider communication in pediatric SCD. Aspects of patient/family-provider communication in pediatric SCD not currently captured by the PCC framework will also be discussed. Findings underscore the importance of family-centered communication across many aspects of SCD care and will inform future research to improve patient/family-provider interactions and health outcomes for this underserved population.

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HYDROXYUREA ADHERENCE THROUGH MOTIVATIONAL INTERVIEWING

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Background: The purpose of the Hydroxyurea Adherence Through Motivational Interviewing (HATMI) study is to identify factors associated with medication adherence, nonadherence, and strategies to improve adherence to Hydroxyurea (HU) from the perspective of persons with sickle cell disease. The research question: In persons with SCD, what is the outcome of motivational interviewing on HU use? The hypothesis: Motivational interviewing will help participants to adhere to HU therapy.

Methods: A prospective cohort pilot study will collect qualitative and quantifiable information on a group of individuals with sickle cell disease. Due to the COVID-19 pandemic all study activities have been carried out using remote communication tools such as a phone or Microsoft Teams®. Subjects were recruited through a southeastern university sickle cell clinic according to institutional review board policy. Inclusion criteria for participation were persons receiving health care from a university sickle cell clinic, diagnosis of sickle cell disease, adults (males and females), ages 18 or older, English-speaking, able to read and write English, currently prescribed HU, experiencing self-identified adherence barriers to HU, and have access to a smartphone and/or computer with internet access. Data were collected by phone or online interviews along with surveys: a health updates form (demographic and medication information), the Patient Reported Outcomes Measurement Information System (PROMIS®) Self-Efficacy for Managing Chronic Conditions – Managing Medications and Treatment Short Form 8a to assess medication adherence behavior, a questionnaire to evaluate satisfaction with the Microsoft Teams® online interviews, a one-dimensional instrument (scale) to assess the subjects readiness to change, and use of a script for semi-structured interviews. The interviews used motivational interviewing techniques, and include the support of managed problem-solving and successful habit-forming behaviors. Objective adherence evaluation was done by calculating medication possession ratio (MPR) based on pictures taken of prescription refill dates by the subjects (per refill) using a personal cell phone and sending the picture(s) to the PI. Data for all meetings will be collected by tape recorded interviews, survey/questionnaires in Research Electronic Data Capture (REDCap), and note-taking. A qualitative approach will be used to document the interview process and analyze the participants' behavior change across the interactions.

Results: The current findings are preliminary and the study is ongoing. All participants self-report as African American/Black. There is an equal distribution among male/female participants. The ages of participants range from 20 – 50 years. Seventy percent have HbSS, 1 participant has HbS thal+, and 2 with unknown sickle cell type. At baseline, the mean score for how ready participants were to change medication taking behaviors was 3.2 on a 10-point scale.

Conclusions: The HATMI study is exploring personal strategies including: managed problem solving, habit forming behaviors, and the effect of motivational interviewing (MI) as an intervention technique to improve adherence to HU medication therapy, a medication known to improve health outcomes in persons with sickle cell disease. The participants will be followed over time to gather data about exposure to and the effect of motivational interviewing (MI). The information acquired from this study will contribute knowledge to the scientific and health care communities and may model interventions that will benefit others with sickle cell disease.

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PSYCHOSOCIAL CORRELATES OF CHRONIC PAIN AMONG YOUTH WITH SICKLE CELL DISEASE

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Background: Approximately 23% of children and adolescents with sickle cell disease (SCD) report chronic pain that can persist into adulthood contributing to poor health-related quality of life and frequent health care use. There is very limited empirical evidence that describes the maintenance of the chronic SCD pain phenotype over time or what biopsychosocial factors may contribute to the development or persistence of chronic SCD pain in youth. The variability in the chronic SCD pain experience requires the need to better understand the interacting individual and family psychosocial factors that may contribute to changes in chronic pain in SCD. This longitudinal cohort study was designed to understand the natural course of chronic pain in youth with SCD and identify potential mechanisms that confer risk or resilience for chronic pain. This study presents baseline assessment data on the cohort to describe pain and psychosocial characteristics of youth with chronic SCD pain and their caregivers. Youth with chronic SCD pain and their caregivers were expected to report poor psychosocial functioning. Worse parent psychosocial functioning was expected to be associated with reduced child functioning.

Methods: Youth receiving care at comprehensive SCD clinics at three tertiary care locations at Children's Healthcare of Atlanta were included if they were aged 10-18 years, any SCD genotype, met the diagnostic criteria for chronic SCD pain (i.e., pain on most days per month, lasting for at least 6 months), and read and spoke English. Youth were excluded if they had comorbid medical conditions typically associated with pain but unrelated to SCD (e.g., rheumatologic disorders or inflammatory bowel disease) or had significant cognitive or developmental limitations that would impair completion of self-report measures. As part of a larger 24-month longitudinal observational study on the natural course of chronic SCD pain, youth and their parents completed a battery of self-report measures at a baseline assessment. Youth reported on pain characteristics and pain-related functioning (Adolescent Pediatric Pain Tool, PROMIS Pediatric Pain Interference) and psychosocial functioning (Pain Catastrophizing Scale – Child; PROMIS Pediatric Anxiety and Depressive Symptoms, Adolescent Sleep Wake Scale). Parents completed self-report measures of parenting stress (Pediatric Inventory for Parents), catastrophic thinking of their child's pain (Pain Catastrophizing Scale – Parent Proxy), and demographics. Descriptive statistics were used to characterize youth pain and functioning and to describe psychosocial functioning of caregivers. Correlational analyses evaluated associations among child and parent psychosocial functioning.

Results: Youth (n=61) were on average 13.9 years old (SD=2.5), 57% female, 95% Black, and 65.5% had HbSS genotype. Parents were on average 39.3 years old (SD=10.7), 92% female, 45.9% single caregiver, and 60% had annual income ≤ $30,000. On average, youth reported a typical pain intensity of 5.1 (SD=2.2), worst pain intensity of 7.2 (SD=2.3), and most described pain quality as hurting (68.9%), annoying (62.3%), awful (60.7%), and throbbing (57.4%). Most youth in this sample reported pain interference of moderate severity (M=61.9, SD=8.8), high levels of catastrophic thinking (M=26.9, SD=11.9), mild depressive symptoms (M=52.9, SD=11.7), and anxiety symptoms (M=47.3, SD=12.3) and sleep quality (M=3.9, SD=0.8) within normative range. Caregivers reported high levels of catastrophic thinking about their child's pain (M=28.9, SD=11.7) and parenting stress within normative range (M=108.9, SD=36.1). Child typical pain intensity was only associated with child pain interference ($r=.31$, $p < .05$). Child pain interference was related to child pain catastrophizing ($r=.57$, $p < .001$), depressive symptoms ($r=.48$, $p < .001$), anxiety symptoms ($r=.37$, $p < .01$), and sleep quality ($r=-.34$, $p < .01$). Caregiver pain catastrophizing was only related to caregiver stress ($r=.66$, $p < .001$). Caregiver stress was concurrently associated with child sleep quality only ($r=-.37$, $p < .05$).

Conclusions: Several aspects of child psychosocial functioning were significantly elevated falling within

moderate to high ranges of clinical risk and associated with child pain-related impairment. Although caregiver stress was not significantly related to child pain or psychosocial functioning, poor child sleep quality was associated with higher caregiver catastrophic thinking about child pain. Chronic pain may exacerbate psychosocial functioning for youth with SCD and caregivers. Results may help identify potential risk factors and parent-child interactions that contribute to the maintenance of chronic pain over time for youth with SCD.

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The effect of the COVID-19 pandemic on caregivers of children with sickle cell disease

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Background: Hydroxyurea (HU) is the primary medication used to prevent the significant medical and neurologic morbidities of pediatric sickle cell disease (SCD; HbSS or HbSB0 thalassemia). Despite the benefits of HU, it remains under-utilized likely due to lack of clinician knowledge/training and negative caregiver perceptions. Thus, we developed the Engage-HU randomized controlled trial (NCT03442114) as a novel approach to address HU utilization barriers. Engage-HU is designed to assess how clinicians can engage caregivers in a shared discussion that considers their values and preferences and scientific evidence about HU. The COVID-19 pandemic has resulted in significant changes to healthcare delivery for children with SCD, as they are at increased risk of severe illness from COVID-19 infection. Concurrently the pandemic resulted in suspending enrolment for most research studies. Beginning in March 2020, Engage-HU study sites stopped enrolling in-person and visits for patients with non-urgent concerns were canceled. Given their risk status, it was recommended that patients with SCD complete telehealth visits when possible. Some families also chose to delay care because they feared their child would get infected at hospitals/healthcare clinics that care for COVID-19 positive patients. Since the lives of all of the families enrolled in the Engage-HU trial have been affected to some extent, we incorporated new measures to capture the impact of the COVID-19 pandemic and the usability of telemedicine implementation and services. We also analyzed whether the impact was related to caregiver age.

Methods: The Engage-HU study aims to recruit 174 caregivers of young patients (0-5 years) with SCD who are considering initiating HU. The trial is being conducted at 9 sites in the United States. Data collection is ongoing and 127 caregiver-participants have been enrolled to date. Since May 2020, caregiver-participants have completed the COVID-19 Exposure and Family Impact Survey (CEFIS), which contains 2 subscales (exposure to potentially traumatic aspects of the pandemic, the impact of the pandemic), and the COVID-19 telemedicine use survey during a baseline study visit.

Results: Currently, 5 of the 9 sites have collected data from 16 caregivers (100% mothers), who all identify as African-American/Black (see Figure 1). Caregivers reported limited COVID-19 exposure to potentially traumatic aspects of the pandemic, yet significant impact of the pandemic on family life. Distress related to COVID-19 varied widely across the sample, for both mothers and children (see Figure 2). Correlations indicated that younger mothers experienced greater exposure to potentially traumatic aspects (r = -.33), whereas older mothers experienced more negative impact of the pandemic (r = .36). Scores on the telemedicine usability survey were generally high, indicating that mothers are happy with the quality of care delivered via telehealth.

Conclusions: Although Engage-HU sites have resumed research operations, recruitment has not reached pre-pandemic targets, as fewer eligible patients are being scheduled for routine care visits at SCD clinics, which accounts for our small sample size. Our preliminary analyses suggest a significant continued impact of the pandemic on families, particularly for those with older caregivers, and general satisfaction with the quality of healthcare delivered via telemedicine.

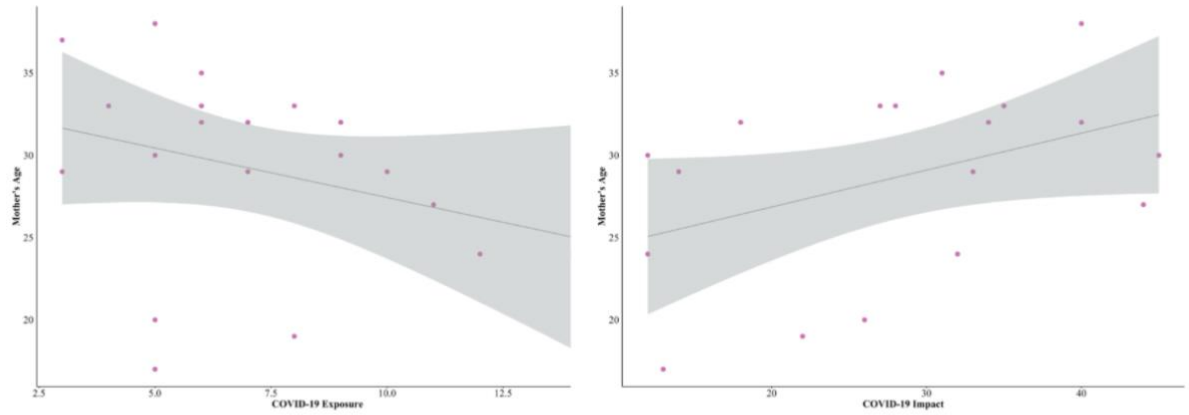

Figure 1. Correlation between mother's age and COVID-19 exposure and impact

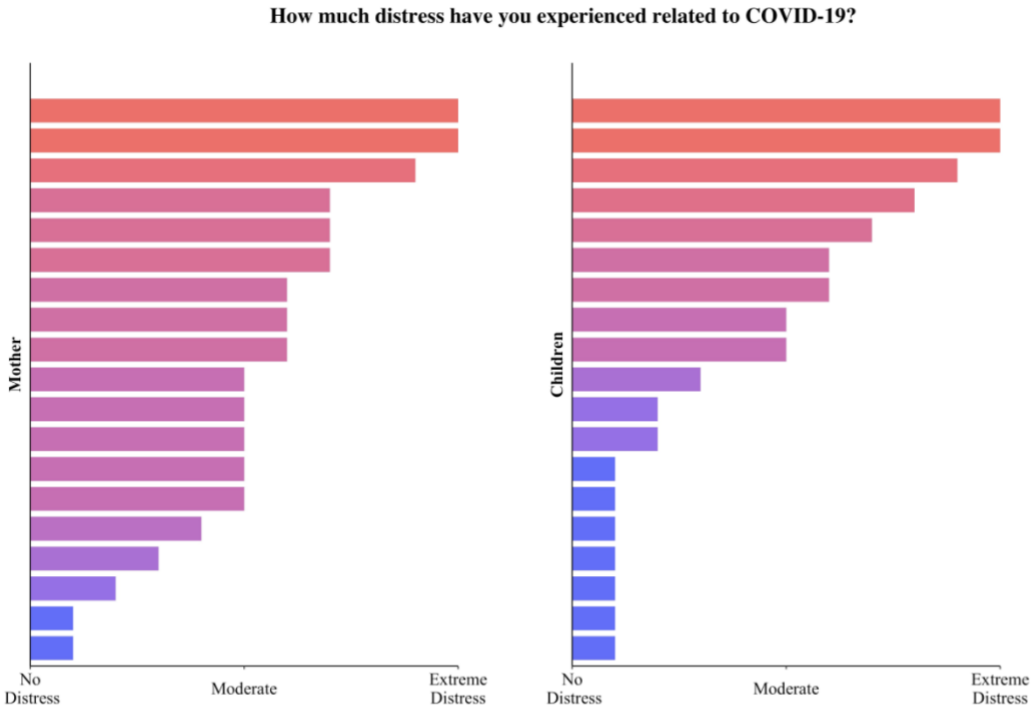

Figure 2. Caregivers were asked how much distress they and their children had experienced related to COVID-19. Each bar represents a response from each caregiver rated on a scale from "No Distress = 0" to "Extreme Distress = 10."

TABLE OF AUTHORS