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Awareness and Use of the Sickle Cell Disease Toolbox by Primary Care Providers in North Carolina

Stephanie O. Ibemere[1], Paula Tanabe[1,2], Emily Bonnabeau[1], Gary Rains[2], Kern Eason[3], Marian F. Earls[4], and Nirmish Shah[2]

Abstract

Background: Sickle cell disease (SCD) is a complex chronic blood disorder characterized by severe disease complications ideally managed by both hematologists and primary care providers (PCP's). PCP's report knowledge gaps and discomfort with SCD management. Our team developed and a decision support tool for SCD management (SCD Toolbox) based on the National Heart, Lung, and Blood Institute's SCD guidelines. We surveyed PCPs in North Carolina (NC) prior to formal dissemination to determine current co-management practices, assess toolbox acceptability, use, format preferences, and understand which algorithms would be most helpful. **Method:** A 23-item baseline needs assessment survey was disseminated to PCPs throughout NC. **Results:** A total of 63 medical providers responded to the survey and of these respondents, 64% reported caring for 1 to 10 patients with SCD. Only 39% of PCPs reported regular communication with an SCD specialist. Providers reported the highest level of awareness of the pediatric and adult health maintenance tools (41% and 39% respectively) and highest use of the pediatric (26%) and adult (28%) health maintenance tools. Respondents also expressed a desire to have access to multiple toolbox formats (37%) (website, mobile app and/or paper). **Limitations:** The use of a convenience sample and low survey response are study limitations which hinder generalizability. **Conclusions:** PCPs rarely co-managed with a specialist, had low awareness and use of SCD toolbox, and requested multiple formats for the toolbox.

Keywords

co-management, sickle cell, primary care, decision support, evidencebased

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Background

Sickle cell disease (SCD) is the most commonly inherited monogenetic hematological disease in the United States.[1] Patients with SCD often experience severe, pain vaso-occlusive episodes, stroke, acute chest syndrome, renal failure, and sepsis.[2] In North Carolina, between 2004 and 2008, the most common complications of SCD were acute chest syndrome and/or pneumonia, renal failure, and anemia.[3] The chronicity of SCD leads to increased morbidity and premature mortality and often results in frequent and costly acute healthcare services.[4-7] Moreover, a high number of emergency department (ED) visits (as many as 6 ED visits/year/person for those aged 18-35 in North Carolina),[3] hospitalizations, and readmissions are associated with the high cost of SCD care.[7-9] However, this increase in cost has not resulted in better health outcomes for patients with SCD.[2] For individuals with commercial insurance and no SCD vaso-occlusive episodes (VOE) in the last 12 months, total annual all cause healthcare costs were $15 747 whereas those with 2 or more VOEs had annual costs totaling $64 555.[10] Total all cause healthcare costs for individuals with public insurance policies (Medicaid and Medicare) who had no VOEs in the last 12 months were $16 750 and $21 877 respectively. In contrast, those with 2 or more VOEs accumulated $$64 566 and $58 308 in total healthcare costs respectively. When stratified by number of VOEs,

[1]Duke University School of Nursing, Durham, NC, USA
[2]Duke University School of Medicine, Durham, NC, USA
[3]Previously Community Care North Carolina, Cary, NC, USA
[4]Marian F. Earls Consulting LLC, Greensboro, NC, USA

Corresponding Author:
Stephanie O. Ibemere, Duke University School of Nursing, 307 Trent Drive, DUMC 3322, Durham, NC 27708-0187, USA.
Email: stephanie.ibemere@duke.edu

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approximately 74.9%, 84.4%, and 95.3% of these total costs were attributed to SCD care when individuals had no, 1 or 2 or more VOEs over 12 months.

Given the severity of this disease, acute care utilization is expected. However, models of care that shift and leverage acute care to the outpatient primary care setting may reduce healthcare utilization and cost encumbered by patients and improve patient health related outcomes.[5] In 2014, the National Heart, Lung, and Blood Institute (NHLBI) developed an evidence-based report and guidelines to facilitate SCD management, particularly for primary care practitioners and other healthcare providers.[11] These guidelines include routine health maintenance guidance as well as recommendations for acute and chronic care management for patients with SCD. Despite this report, recent research of primary care providers suggests a general discomfort with the management of SCD.[12] This discomfort stems not only from the complexity of SCD and the medications used to modify the disease, but also from provider reported knowledge gaps related to SCD management.[13,14] However, these primary care providers did note they would be amenable to working with SCD/hematology specialists to manage the care of their patients with SCD. These findings provided an opportunity to increase provider awareness and engagement with evidence-based practice recommendations for SCD treatment and management.[12,14]

In order to improve care provided to patients with SCD, we partnered with Community Care North Carolina network (CCNC) and regional experts to develop and begin a small, informal dissemination of evidence-based guidelines for the treatment of SCD.[15] During this study, CCNC operated as a Medicaid care management program focused on improving primary care access and quality. Prior to statewide dissemination efforts and following our informal dissemination to focused practices, we subsequently aimed to assess co-management practices, as well as awareness and use of the toolbox.

Methods

Study Design

To assess PCP baseline awareness and use of the decision support toolbox, and co-management strategies we used a cross-sectional, survey methodology. The Duke University Medical Center institutional review board (IRB) reviewed and approved this study. We developed the SCD Toolbox and aimed to obtain early feedback as to which algorithms and formats would be most useful.

Setting and Sample

The setting for this study has previously been described.[15] In summary, CCNC served as the Medicaid managed care organization for the state of North Carolina. This system included 14 PCP networks across the state. These networks provided care to more than 1 million Medicaid enrollees, including 4392 enrollees diagnosed with SCD between 2016 and 2019. All fourteen networks of primary care providers and pediatricians were invited to participate in the survey. Leadership at CCNC in partnership with practice managers identified the practices in North Carolina with the most patients with SCD.

SCD Toolbox Development

The year prior to our informal dissemination of the toolbox, SCD experts in NC partnered with CCNC leaders, PCPs, pediatricians, and emergency providers to develop paper, web and app based versions of algorithm based decision support tools based on the 2014 NHLBI SCD treatment recommendations. The toolbox included the following decision support algorithms based on the following NHLBI recommendations: routine health maintenance, co-management for hydroxyurea, reproductive counseling, pain, fever, and anemia, respiratory and neurological symptoms. A detailed description of the development of the SCD Toolbox has been reported.[15]

PCP Survey Development and Dissemination

Surveys for PCP's were developed with input from stakeholders including healthcare providers, SCD experts, and researchers. PCP surveys focused on provision of care to patients with SCD, co-management with SCD specialists, as well as awareness, use, and preference for the decision support tools in the SCD toolbox. Survey questions focused on 3 areas: demographic information, clinical practice characteristics, as well as awareness and use of evidence-based decision support tools. Questions focused on clinical practice included yes/no questions which when answered in the affirmative, led participants to answer additional yes/no or Likert-style questions.

Identified practices received letters detailing the study aims and provided the PCP surveys in paper or electronic format via a weblink. Interested providers volunteered to complete the survey on paper or electronically. Paper based responses were returned to the study coordinating center at Duke University via email or gathered locally by CCNC QI specialists for pick up by Duke study staff.

Data Analysis

Descriptive statistics were used to characterize all survey data. Dichotomous and Likert-style responses have been represented as frequencies and percentages. All statistical analyses were conducted using SAS 9.4 (SAS Institute, Cary, NC).

Results

Surveys were disseminated to 463 PCPs across 50 PCP practices within 9 different PCP networks. A total of 65 survey responses were received yielding a response rate of 14%. Two survey responses were eliminated as: (1) 1 participant reported their practice area as emergency medicine, (2) and 1 survey was submitted with no responses in any fields; therefore, a total of 63 responses were included in this analysis. Table 1 provides demographic information about the PCPs who responded to these surveys. Of the 37 PCPs reporting their practice area as family practice, 11 respondents primarily cared for adults and 26 cared for adults and children. The second largest group of respondents (n = 20) selected pediatrician as their practice area.

Information regarding PCP practice characteristics and co-management is described in Table 2. The majority of the PCPs (90%) who responded to this survey were in family or pediatric practices with nearly 45% reporting practicing in a rural setting. Two-thirds of this sample also indicated practicing in a private practice setting compared to 21% practicing in an academic setting. Of those PCPs who communicated regularly with an SCD specialist (n = 18), 14 (77.78%) communicated with the SCD specialist sometimes or often while 4 (22.22%) only rarely communicated. Of those respondents who reported they regularly communicate with an SCD specialist, 16 are primary care clinicians for patients with SCD in their practices.

Descriptive statistics related to PCP tool awareness, use of the toolbox, and preferred tool format are displayed in Table 3. There was generally low awareness of the NHLBI recommendations and use of the decision support tools reported by PCPs. While it was expected that the decision support tools related to pediatric fever and health maintenance would be used by PCPs in pediatrics, PCPs in family practice accounted for at least half the number of clinicians using these tools (pediatric fever, n = 5 (50%); pediatric health maintenance, n = 11 (64.71%)). Only 2 pediatricians reported use of the hydroxyurea decision support tool.

Table 1. Primary Care Providers Demographic Information.

Demographics N (%)	(n = 63)
Age, mean (SD)	42.75 (10.69)
Years in practice, mean (SD)	12.68 (10.24)
Sex	
Male	47 (73.44)
Female	16 (25.00)
Race	
African American/Black	17 (26.56)
Asian	3 (4.69)
More than one race	1 (1.56)
White	42 (65.63)
Ethnicity	
Hispanic	1 (1.59)
Non-Hispanic/Latino	59 (93.65)
Unknown/not reported	3 (4.76)
Professional training	
MD	33 (52.38)
PA	8 (12.70)
NP	19 (30.16)
RN	3 (4.76)
Practice area	
Family practice	37 (58.73)
Internist	1 (1.59)
Pediatrician	20 (31.75)
Other	5 (7.95)
Geographic location of majority of providers' patients	
Urban	14 (22.58)
Suburban	21 (33.87)
Rural	27 (43.55)
Unknown/not reported	1
Practice setting	
Community hospital	7 (11.67)
Private practice	40 (66.67)
Academic setting	13 (21.67)
Unknown/not reported	3
Primary patient population	
Children	21 (33.33)
Adult	13 (20.63)
Both	29 (46.03)

Practice area category "other" for baseline data included participant responses of adult primary care (n = 1), internal medicine and pediatrics (n = 3), pediatric nurse (n = 1).

Discussion

Low awareness and use of the SCD toolbox was not surprising as we distributed this survey prior to a formal statewide dissemination. However, some respondents were aware of the tools. This can be explained by a very small, informal word of mouth dissemination of the SCD toolbox that occurred prior to survey distribution by several hematologists with a strong interest in SCD who shared these tools informally. *The purpose of this small informal dissemination was to introduce the toolbox and obtain early feedback which could guide future and more formal dissemination efforts.*

Our findings indicated few PCPs regularly communicated with SCD specialists and ultimately, had low awareness and use of the toolbox. Notably, participants in the current study did not report large panels of patients with SCD, which has been associated with decreased comfort with SCD management for PCPs.[12,14] Considering the relatively small patient panels reported in the current study and limited frequency with which PCPs provided care to a person with SCD, discomfort and low use of the SCD toolbox

Table 2. Practice Characteristics and Co-Management.

Survey questions	(n = 63), n (%)
Sickle cell disease care and co-management	Yes
Are you a primary care clinician for patients with SCD in your practice?	32 (53.33)
Have you ever cared for a patient with SCD?	49 (77.78)
In the past year, have you cared for a patient with SCD?	30 (63.83)
Do you regularly communicate with the sickle cell specialist who cares for your patients with SCD?	18 (39.13)
How many patients do you follow with SCD?	
None	14 (29.17)
1-10	31 (64.58)
11 or more	3 (6.24)

Missing data are not included in the total count nor percentage for these items.

Table 3. Primary Care Provider Awareness, Use of, and Preference for SCD Decision Support Tools.

Survey questions	(n = 63), n (%)	
Decision support tools	Tool awareness	Tool use
Adult fever	10 (15.87)	5 (7.81)
Pediatric fever	16 (25.40)	10 (15.87)
Adult health maintenance	26 (41.27)	18 (28.57)
Pediatric health maintenance	25 (39.68)	17 (26.98)
Anemia	12 (19.05)	7 (11.11)
Hydroxyurea	10 (15.87)	6 (9.52)
Neurological symptoms	4 (6.35)	0 (100)
Pain management	17 (26.98)	10 (15.87)
Respiratory symptoms	13 (20.63)	5 (7.94)
Tool preference		
Website	18 (33.96)	
Mobile app	10 (18.87)	
Paper	5 (9.43)	
All of the above	20 (37.74)	

Those who did not report a preference are not included in the total count nor the percentages for this item.

is not surprising. Furthermore, although co-management for chronic disease management can improve patient outcomes,[16,17] uptake of co-management for SCD has been slow.[5]

Approximately half of our study sample provided care to a person living with SCD and among this group only 39% co-managed care with their patient's SCD specialist. Though the results of these studies show PCPs were amenable to co-management with an SCD expert, the participants of this study did not report high levels of co-management. An analysis of 8 health systems indicated those patients co-managed by a PCP and a hematologist were less likely to be hospitalized frequently (defined as hospitalized more than once a year) when compared to patients who were not co-managed.[18] Moreover, there is evidence to suggest patients and providers would like to adopt a co-management model for SCD care, however, implementation has been limited due to poor access to specialist, poor communication and lack of medical record integration and knowledge gaps.[12,14,19,20] Clinical decision support tools have been proposed as a potential solution to these issues[12,21] and the SCD toolbox is equipped to provide pertinent clinical information PCPs can utilize to maintain the health of their patients with SCD while providing access to SCD specialists.[22] It is evident, there is an opportunity to increase co-management of patients with SCD in NC.

Participants of the current study expressed higher levels of awareness with adult and pediatric health maintenance, pediatric fever, and pain management tools in the SCD Toolbox. However, the participants aware of these tools only accounted for less than 1/3 of the study sample. The results of this study are similar to previous reports of low PCP awareness of the NHLBI guidelines for SCD management.[12,14,22] As noted in these studies, many PCPs are unaware of these guidelines and expressed general discomfort with SCD management due to the complexity of disease management. The extant evidence

suggests PCPs were also inclined to utilize clinical decision support for SCD management, however, participants of this study reported low levels of use of the SCD toolbox.[12,22] The information garnered from this study offers insight on the types of SCD-related decision support tools which are most helpful, specifically the use of pediatric and adult health maintenance tools in primary and family practice care. Use of the SCD toolbox aims to increase the quality of care which PCPs can offer to patients with SCD by connecting PCPs to SCD experts and providing strategies to prevent morbidity and premature mortality in the primary care setting. For those that wish to review the tools available, the SCD Toolbox remains readily available and can be accessed through this link: https://www.scdtoolbox.com/.

Preference for decision support format, however, showed participants equally appreciated having decision support online, in paper, and through a phone app interface. These data demonstrate the importance of tool accessibility as format preferences can vary not only by PCP preference, but also resource availability and the demands of outpatient clinical care. These different tool formats allow for the SCD Toolbox to be integrated into local electronic health systems but also easily accessed through a smart phone application or via paper for quick reference during busy clinic days. These formats can enhance the PCP's ability to identify, evaluate, and mitigate risk factors associated with acute and chronic complications of SCD in the primary care setting. Moreover, the multiple ways in which the SCD Toolbox can be implemented can reduce barriers related to knowledge, location (urban versus rural), local access to SCD experts, or practice setting (academic versus community practice). Given the infrastructure already available through the North Carolina Sickle Cell Syndrome program (supported by the North Carolina Department of Health and Human Services), further dissemination of the SCD Toolbox is possible.[23] Considering a prominent goal of this program is to reduce morbidity and mortality associated SCD, these decision support tools could further support this mission and provide a central home which is accessible to both providers and patients. Moreover, the SCD Toolbox could be integrated into the training this program already provides to healthcare professionals.

Limitations

These results should be considered within the context of the following study limitations. The response rate to the survey was low and provided a small sample from which to draw conclusions. The information gathered may therefore not be representative of the larger population of PCPs in North Carolina given the sample size. However, the survey provides a snapshot understanding of co-management practices and identifies elements of the toolbox which may be most beneficial, as well as the need for multiple modes of availability.

Conclusions

The findings of this study suggest PCPs found several elements of the toolbox helpful and indicated they would benefit from multiple different formats of the tools. Future efforts will require wider dissemination efforts to increase awareness and use of the toolbox.

Declaration of Conflicting Interests

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ORCID iD

Stephanie O. Ibemere https://orcid.org/0000-0002-2006-9924

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