Healthcare Utilization and the Quality of Life of Children and Adolescents with Sickle Cell Disease

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Abstract

Introduction: Youth diagnosed with sickle cell disease (SCD) often utilize the healthcare setting to manage disease-related symptoms, yet the connection between healthcare use and their overall wellbeing has been understudied. This study investigates whether healthcare utilization predicts the health-related quality of life (HRQOL) in youth with SCD. It is hypothesized that increased healthcare utilization will predict lower HRQOL in pediatric SCD. Methods: A total of 150 patients, ages 8-17 years old, were enrolled in this cross-sectional quantitative study. Patients completed the Pediatric Quality of Life (PedsQL) 3.0 SCD module, while the researcher conducted a retrospective chart review to gather patient characteristics such as emergency room (ER) and hospitalization occurrences over the past 12 months. Results: A higher frequency of ER visits ($p < .05$) and hospitalizations ($p < .01$) predicted lower HRQOL scores. Age ($p < .05$) also emerged as a significant predictor for both regression models. Discussion: As youth with SCD require ER treatment and/or hospital admission, they are at increased risk for lower HRQOL, specifically as they get older.

INTRODUCTION

Sickle cell disease (SCD) is a genetic, chronic blood disorder that is characterized by the functioning of red blood cells (RBC). Normal RBCs are circular in shape, which allows the cell to carry out tasked such as tissue oxygenation, amongst other vital responsibilities (Conran & Embury, 2021). Sickled cells, however, take on a C-shape, which leads to a blockage of the capillaries causing inflammation and severe pain crises (Knisely et al., 2020). In addition to pain, individuals with SCD are at risk for chronic organ damage, anemia, and early death (Wallen et al., 2014).

Pain is often considered the hallmark symptom associated with SCD; therefore, researchers have attempted to understand the pain experience among individuals within this population. The Pain in Sickle Cell Epidemiology Study (PiSCES) highlights this experience and reports that patients will typically manage their pain at home to avoid going to a busy ER (Smith et al., 2005). While the pain experience, which has been shown to increase with age, has been linked to mental health challenges such as depression and anxiety (Valrie et al., 2020), it is also the most common reason patients access the hospital (Kanter et al., 2019).

Healthcare utilization among individuals with SCD is increasing along with the associated costs. One study found the annual cost of managing individual with SCD ranged from \$18,859 for ages 0 – 9 to \$43,586 for ages 20 – 29 annually (Salcedo et al., 2019). In a different study, SCD accounted for one of the highest rates for readmission among chronic illnesses and accounted for approximately \$2.4 billion dollars annually (Crego et al., 2020). While the economic burden associated with SCD is both an individual and national concern, high healthcare utilization also has psychosocial implications. Kidwell et al. (2021), for example, found depression and anxiety scores were higher among individuals who accessed the ED and were admitted to the hospital more frequent. How patients cope with the many stressors associated with SCD is vital, as it may determine their overall wellbeing.

Health-related quality of life (HRQOL) is a self-assessment of a patient's overall health as it relates to their disease status (Theofilou, 2013). The literature has been consistent in reporting that children diagnosed with SCD have poorer HRQOL even when compared with children without SCD (Menezes et al., 2013; Wrotniak et al., 2014). However, it is difficult to generalize these findings for two primary reasons. First, researchers in this field have used various measurement tools to assess HRQOL within this population. Also, studies have neglected using a disease-specific measurement tool when assessing HRQOL. Those interested in this type of research may find it beneficial to use a disease-specific measurement tool that consist of domains specifically related to SCD. Secondly, many research studies have omitted using a theoretical model to guide their study and/or substantiate their findings. One may find it useful to include a theoretical model to better understand the connection between youth with SCD and HRQOL.

One such theoretical model that illustrates the connection between healthcare utilization and the HRQOL of youth with SCD is the illness intrusiveness theory. Developed by Devins et al. (1990), the illness intrusiveness theory posits disease, treatment, psychological, social, and contextual factors cause illness intrusiveness, which affects the wellbeing of individuals with chronic illness (Devins, 2006). Patients who perceive their illness to be intrusive on their daily functions and activities they enjoy report lower HRQOL (Devins et al., 2000), even within children diagnosed with a chronic illness (Fedele et al., 2012). As children with SCD continue to access the emergency room and/or require admission to the hospital, they are unable to engage in positive experiences, which are vital during child and adolescent years (Devins et al., 1993). Prior research has used illness intrusiveness as a theoretical framework within the African American population (Bioku et al., 2020; Hughes et al., 2014), which underscores the value in using this theory for the current study. Because symptoms such as pain often leads to emergency room visits and hospital admission, it may be likely that the way individuals adjust and are able to decrease the need to access the hospital will aid in improving their overall HRQOL. To discover if there is a connection between healthcare use and the HRQOL of youth with SCD, this study will use the Pediatric Quality of Life Scale (PedsQL) 3.0 SCD module to build on the literature. There are two identified aims: (1) to investigate whether youth diagnosed with SCD experience low HRQOL using the PedsQL 3.0 and (2) analyze the association between healthcare use and the HRQOL of children and adolescents diagnosed with SCD while also controlling for specific sociodemographic variables. It is hypothesized that there will be an impairment in HRQOL and there will be a correlation between healthcare utilization and the HRQOL of youth with SCD. Findings from this study may aid healthcare professionals in incorporating mental and emotional support in their treatment of children with SCD.

METHODOLOGY

The purpose of this study is to investigate whether the HRQOL of children and adolescents with SCD is impaired and to examine the association between healthcare utilization and the HRQOL scores. Therefore, this study deployed a cross-sectional quantitative research study to analyze these variables. The current study was approved by the Institutional Review Boards (IRB) of Eastern Virginia Medical School and Norfolk State University.

Participants and Procedure

Research participants were recruited from a comprehensive sickle cell program at a local children's hospital in Southeastern Virginia between July 2018 – February 2019. The inclusion criteria required patients and their parents to speak and read English fluently. For the patients specifically, the inclusion criteria were as follows: have a current diagnosis of SCD, be between the ages of 8 – 17 years old, and to not have been diagnosed with any comorbid developmental disabilities. Parents had to be at least 18 years old and identify themselves as the legal guardian. Caregivers who were not the biological parents, but had legal guardianship were allowed to participate in this study. For this reason, the word "parent" and "caregiver" are used interchangeably. Parents and caregivers were excluded from participating this study if they did not meet the inclusion criteria.

As families agreed to participate in the study, patients signed assent and parents signed consent forms. Afterwards, patients were asked to complete the Pediatric Quality of Life Scale (PedsQL) 3.0 SCD question-

naire, while parents completed a demographic information questionnaire. Lastly, as families completed the questionnaires, the recruiter reviewed the documents for completeness and proceeded to review the patient's electronic medical record (EMR) for the retrospective chart review. A full description of the PedsQL 3.0 SCD module, demographic information questionnaire, and chart review are provided further in the document.

Measures

Dependent Variable

The PedsQL 3.0 SCD module, a disease-specific measurement tool, was specifically selected for this research study to examine the HRQOL of the participants. There are two separate questionnaires used – one for our child participants between 8 - 12 years old and one for our teen participants 13 -17 years old. Both questionnaires contain nine different dimensions, 43 items total, with each dimension assessing the patient's pain, worry, emotions, treatment, or communication. The total score for the scale, which are reversed scored, was calculated by using the sum of all items divided by the number of items answered. Lower scores on the questionnaire suggests more problems related to HRQOL. The Cronbach's alpha score (.95) shows good reliability and intercorrelations with PedsQL Multidimensional Fatigue Scales and PedsQL Generic Core Scales reported medium to large construct validity with scores between .30 and .50 (Panepinto et al., 2013).

Independent Variable

A retrospective chart review of the patient's EMR was conducted to gather information related to healthcare utilization. After the patient completed the PedsQL 3.0 SCD module, how often the patient accessed the ED and the frequency of hospitalizations over the previous 12 months were recorded. This strategy has been used in previous studies (Kidwell et al., 2021).

Covariates

Parents and guardians were asked to complete a demographic information questionnaire, which contains information related to the patient and parent. This form asked for information such as the patient's age and gender. It also asked for the caregiver's information including their highest level of education completed, marital status, and household income. The information gathered from this demographic sheet have been known to cause variance in HRQOL studies and have been obtained in previous studies (Abdallah et al., 2020); therefore, it was important to obtain.

Data Analysis

The most recent version of the Statistical Product and Service Solutions (SPSS) package was used for data analysis. First, a univariate analysis was conducted on the PedsQL 3.0 SCD module, the frequency of healthcare use (ER and hospitalizations) for patients over the previous 12 months, and sociodemographic information for patient. Multiple Regression analysis was used to examine the association between HRQOL and the healthcare utilization among youth with SCD while controlling for sociodemographic variables. Categorical variables were transformed to dichotomous variables for this analysis.

RESULTS

A total of 150 patients were enrolled in the current study. Table 1 provides a description of the demographic information from the study sample. The mean age of the participants was 12 years old, while most patients in the study identified as female (52%).

Pediatric Quality of Life Scale 3.0 SCD Module

Table II provides an overview of the descriptive analysis of the PedsQL 3.0 SCD module and nine domains. The mean score of the PedsQL 3.0 SCD module fell in the middle of the reference distribution ($M = 59.36$, SD = 19.58). Regarding the subscales, the lowest reported mean score was in response to *communication II* ($M = 45.22$, SD = 29.30), suggesting that respondents struggled most with communicating with others about having SCD. Examining the pain categories, respondents scored the lowest in the area of *pain impact*

(M = 49.48, SD = 26.15), which implies that, as participants experienced painful events, it significantly disrupts their daily functioning.

Healthcare Utilization

A retrospective chart review (Table III) was conducted to examine the frequency of ER visits and hospitalizations for each patient over the past year. Only a small number of patients frequented the ER (7%) and were hospitalized (5%) [?]4 times over the past year, which is above the average number of visits per year for this patient population (Center for Disease Control and Prevention [CDC], 2020) and has been the cutoff to distinguish between low and high healthcare utilizers in prior research (Paulukonis et al., 2017).

Multiple Regression

To examine whether healthcare utilization is a predictor of HRQOL in youth with SCD, multiple regression analysis was deployed. There was a statistically significant relationship observed between HRQOL and the frequency of ER visits (B = -1.05, P = .04) and hospitalizations (B = -2.73, P = .003) over the past 12 months (Table IV). There was also a relationship observed between age and HRQOL in both regression models (P = .03).

DISCUSSION

This study examined the HRQOL of children and adolescents diagnosed with SCD using a disease-specific measurement tool and to investigate the relationship between healthcare utilization and HRQOL in this population. Comparing the results of this study with previous research (Beverung et al., 2015), it was discovered that youth in this sample experienced impairment in their HRQOL. This information mirrors the results found in the literature. Wrotniak et al., 2014, for example, studied compared HRQOL scores of children with SCD with normative scores of non-White children and found that the scores of children with SCD were lower. Similar results were even noted in studies where the HRQOL of children with SCD were compared with children diagnosed with other chronic illness (Bhagat et al., 2014). The complex nature of SCD makes it difficult to pinpoint a sole culprit for impairment in HRQOL; rather, there have been many biopsychosocial factors that have been associated with their wellbeing such as disease severity, depression, anxiety, and parenting stress (Barakat et al., 2008; Goldstein-Leever et al., 2020).

Participants in this study noted communication to be the most challenging. This was not unusual and, in fact, other studies have reported how youth are cautious about disclosing their diagnosis of SCD to others. Many children with SCD do not want to continuously explain their illness to others and some even feel emotional, as it reminds them of the hardship they face managing their disease (Forrester et al., 2015).

Concerning the relationship between healthcare utilization and HRQOL, this study found that the frequency in healthcare utilization predicted HRQOL scores, suggesting that the more patients accessed the ER or were admitted to the hospital, they may expect to have lower HRQOL. This finding is consistent with previous, but scant, literature that examined healthcare utilization and HRQOL among youth with SCD (Ahmed et al., 2016). Interestingly, research on this topic within other chronic illness groups such as cancer (Choo et al., 2019) and arthritis (Moorthy et al., 2010) have comparable results. In SCD specifically, prior research has shown pain to be the main reason for hospitalizations and ER visits (Cacciotti et al., 2017; Kidwell et al., 2021), and has been linked to decreased HRQOL (Badawy et al.,2018). Moreover, research has shown that the frequency and intensity of pain increases in severity as children grow older (Zempsky et al., 2016). Perhaps this rise in pain severity as children age is linked to the connection between age and healthcare utilization. This association has been found in previous studies (Fosdal & Wojner-Alexandrov, 2007; Panepinto et al., 2005) and was a significant predictor for HRQOL in the current study.

LIMITATIONS

There are several limitations that are worth mentioning. Concerning the HRQOL variable, this data captured the patient's experience within the past month. While this method was most appropriate for this study

design, examining HRQOL over a broader period may depict a deeper understanding of the connection between healthcare utilization and HRQOL.

In referencing the inclusion/exclusion criteria, this study excluded patients with comorbid developmental disabilities, but did not exclude patients with comorbid physical illnesses. The PedsQL 3.0 assisted with gathering data related to SCD as opposed to using a generic scale; however, comorbid diseases such as diabetes may also influence symptoms of SCD that contributes to the frequency of healthcare use. Excluding or controlling for this data may improve the power of this type of study.

While the number of participants included in this study mirrors previous research in this area, the number of patients in the current study is hardly generalizable. The results highlight concerns surrounding healthcare usage and patient wellbeing; however, these results should be interpreted with caution. Perhaps a multicenter, longitudinal study design would further substantiate research in this area.

CONCLUSIONS

To summarize, youth with SCD face insurmountable disease-related symptoms that often disrupts their ability to cope and engage enjoyable activities. The current study found that the frequency in which children and adolescents access the ER and/or require hospitalization predicts their overall HRQOL. Further research may explore interventions that assist patients in learning to cope with symptoms and to decrease their need to access the hospital emergently.

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