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Education Needs of Adults Living with Sickle Cell Disease in the U.S. and Jamaica

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EDUCATION NEEDS OF ADULTS LIVING WITH SICKLE CELL DISEASE IN THE U.S. AND JAMAICA

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ABSTRACT

Purpose: To identify psychosocial and physiological components of sickle cell disease (SCD) health promotion to be included in patient and family education materials provided to an adult living with SCD for the promotion of wellness and reduction in complications of the disease.

Background and significance: SCD occurs in approximately one in five hundred Jamaican born people and about 0.49 out of 1000 in America and results in intense pain episodes, and multi-organ failure (Knight-Madden, 2011). The role of the nurse and other healthcare providers as well as community based organizations in patient education is critical for promotion of quality of life. Tannahill (2009) argues that "fostering of empowering attributes such as resilience, self-esteem, confidence and lifeskills" also falls under the realm of education.

Methods: A combination of telephone and written questionnaires were completed by nurse experts caring for patients with SCD in the U.S. and Jamaica, as well as executive board members of community based organizations for the support of community members living with SCD and their families.

Discussion: There is a consensus that individuals with SCD are not adequately educated about the condition and its implications. Respondents recommended that those living with SCD be taught about basic physiology of the disease, family planning, early signs of complications, and exacerbating factors. Furthermore, each respondent explored barriers to effective educational interventions as well as methods of enhancing the quality and effectiveness of these interactions. This information can be used to develop effective educational materials for improving health outcomes.

BACKGROUND AND SIGNIFICANCE

Sickle cell disease (SCD) is not one, but a category of hemoglobinopathies that affect the oxygen carrying capacity of the blood, as well as blood flow itself. "SCD is the most common genetic condition worldwide (National Heart, Lung, and Blood Institute, 2002)" and "SCD occurs in about 1 in every 500 African American births" (Odesina, Bellni, Leger, Bona, Delaney, & Andemariam, 2010; Valente, Alexander, Blount, Fair, Goldsmith, & Williams, 2010). Approximately one in five hundred Jamaican born people will have some form of SCD (Knight-Madden, 2011). Because of the high prevalence of this disease worldwide, it is necessary to promote the health and wellness of this population to prevent complications and hospitalizations. Despite such prevalence, in a study by Treadwell, McClough, and Vichinsky (2006) only 13.1% of community respondents correctly defined SCD manifestations. Alavi and Kirsner (2015) recognize the growing prevalence of SCD as a public health problem.

Furthermore, SCD can be recognized as an *acute on chronic* condition. By this it is meant that between acute exacerbations, individuals are living with the adverse effects of prior exacerbations throughout the lifespan. Long term sequelae of microvascular occlusion such as chronic pain syndrome, acute chest syndrome, and multi-organ damage warrant targeted health promotion education. According to Platt and Sacerdote (2006, p. 12), chronic complications of SCD include, "moderate to severe anemia, increased infections, tissue damage, organ damage, and recurrent pain episodes." By providing exceptional education, self-efficacy among those living with SCD is increased leading to decreased pain and fewer self-reported symptoms (Clay & Telfair, 2007).

According to Bhagat et al. (2014), SCD follows an autosomal recessive pattern of inheritance, and is characterized by anemia, vascular occlusion, pain, organ failure, and death in

early adulthood, resulting from the production of abnormal hemoglobin, hemoglobin S. In conditions of hypoxia, dehydration, or acidosis, the red blood cells (RBC) elongate, creating a sickle shape, and stick to eachother – the mainstay of SCD (Knight-Madden, Lewis, Tyson, Reid, & MooSang, 2009). The RBC's adhere to the endothelium of capillaries, injuring these tiny blood vessels and promoting vascular inflammation as well as impeding perfusion. The resultant vaso-occlusive events (VOE) cause pain, fever, swelling, strokes, multiple organ failures, leg ulcers, and pain events (Bhagat, Baviskar, Mudey, & Goyal, 2014; Hinkle & Cheever, 2014, p. 909). Autosplenectomy, accounted for by the lysis of the spleen due to microinfaction, in childhood accounts for the heightened risk of infection across the lifespan. Chronic anemia occurs as abnormally shaped erythrocytes become fragile and are rapidly hemolyzed; the average life span of an RBC containing HbS is only ten to twenty days, compared to 120 for a healthy RBC (Valente et al., 2010) . The result is a baseline hemoglobin of 7-10 g/dL, below the low normal range for an adult, leading to cardiomegaly and heart failure among other cardiac complications (Hinkle & Cheever, 2014, p. 909-910). Acute chest syndrome, a complication in which the gas exchanging microvasculature of the lungs is occluded by sickled erythrocytes, exacerbates hypoxemic hypoxia. Many young and middle aged adults living with SCD require the support of portable oxygen systems.

Platt and Sacerdote (2006, p. 12) list some genotypes that include the HbS gene, and thus produce varying degrees of sickle symptoms: HbSS, HbSC, and HbS-beta thalassemia. There are others, however these are most common worldwide. When a child inherits two sickle cell genes from his or her parents, the result is the genotype HbSS – the resultant phenotype is sickle cell anemia. The adult living with HbSS sickle cell anemia experiences life-long pain and tissue death related to abnormal red blood cells occluding vessels supplying oxygen to those tissues.

Childhood micro infarctions commonly result in stroke, blindness, splenic inefficiency, and lung complications. Following from issues related to spleen function, it can be expected that those with this condition will be susceptible to infection across the lifespan. HbSS is the most severe form of the disease. Furthermore, and relevant to this article, strokes in childhood can affect how the individual will learn best when he or she becomes and adult, as well as comprehension and executive functioning, perhaps affecting his or her knowledge related to this condition. Deficient knowledge and impaired organizational planning can hamper adequate self-care. HbSC is a milder form of HbSS, with a similar disease profile. Complications typically occur later in life (Platt & Sacerdote, 2006, p. 13).

A particularly dramatic complication of SCD is sudden cardiac death associated with rhabdomyolysis. This can occur in individuals with the HbS trait (HbAS, or HbSS). In the setting of intense exercise or exertion, acidosis and dehydration as well as localized hypoxia promote the sickling of HbS. The sickled HbS molecules adhere to other HbS molecules and the endothelium of small blood vessels worsening the hypoxic state. Muscular demand for oxygen continues to rise as microvascular occlusion occurs. In this ischemic state, myocytes begin to lyse and intracellular potassium spills into the bloodstream. The sickled RBCs may also leak potassium extracellularly. A sudden rise in serum potassium causes fatal cardiac dysrhythmias and sudden death (Loosemore, Walsh, Morris, Steward, Porter, & Montgomery, 2012). Platt and Sacerdote (2006, p. 52) acknowledge that in a "military study of 2 million recruits, a 28 fold increase in unexplained exercise related deaths occurred in those with sickle cell trait, compared to similar age, sex, and race matched non sickle cell trait recruits." In half of these deaths, sickle cell trait was the only detectable cause (Platt & Sacerdote, 2006, p 52).

In 2007, the National Athletic Trainers' Association released a consensus statement regarding sickle cell trait and the athlete (2007). A multidisciplinary task force firmly recommended that all that all collegiate level student athletes be aware of their SCT status and that coaches and medical staff understand how to recognize and treat sickle cell collapse, which occurs during intense exertion. Sickle cell collapse is characterized by small blood vessels becoming obstructed by thrombi formed when sickled RBC's clump together during hypoxemia, acidosis, or dehydration. As the athlete continues to exercise, muscles become starved of oxygen leading to explosive rhabdomyolysis within the first few minutes of intense exercise (National Athletic Trainers' Association, 2007).

Community based organizations promote the health and wellness of community members living with chronic conditions. A study by Treadwell, McClough, and Vichinsky (2006) shows that "there is limited visibility of and knowledge about SCD and SCT [sickle cell trait] in the general population" and "providers bear responsibility for the community's lack of awareness about SCD and providers needed to improve in educating their individual patients about SCD and SCT." Social support has been shown to increase self-care efficacy and improve outcomes in those living with SCD (Matthie, Jenerette, & McMillan, 2015). Participants older than 33 years of age, in a study by Treadwell and colleagues (2006), were at an increased likelihood of gaining educational information about SCD and SCT from community organizations. In the same study, those who scored highest on knowledge tests regarding SCD and SCT received information from community organizations (Treadwell et al., 2006). This supports the inclusion of responses from executive board members of CBO's in this pilot study.

The intrinsic role of interprofessional health care team is advocacy for the patient. Through patient education, the nurse empowers the patient to be able to advocate for his or

herself enabling autonomy on the part of the patient. Hines, Mitchell, Crosby, Johnson, Valunzuela, Kalinyak, and Joiner (2011), have identified education of patients and families about SCD as a key variable in improving health outcomes. Wright and Adeosun (2009) explain, "Failure to involve patients with SCD in making decisions about their care undermines self-reliance and self-knowledge." When speaking on the literature revolving around adults living with SCD, Abuateya, Atkin, Culley, Dyson, S., and Dyson S., (2008) identify that "many studies, in keeping with their initial focus, … recommend further research on patient education." How is it possible to educate the patients, as Wright and Adeosun (2009) suggest, when there is only limited evidence by which to base the education? SCD is a multifaceted disease with factors including developmental, psychosocial, and sociocultural in addition to the physiological. It is important for nurses to understand in what areas those living with SCD, and their family members, need to be educated. Current research regarding the content of patient educational materials in SCD is sparse. Tanyi (2003) identifies a lack of examination of SCD health promotion, prevention, and maintenance, although "the unpredictable trajectory of SCD can lead to frustration, fear, helplessness, hopelessness, and emotional distress."

METHODS

Participants

Participants in this pilot study include nurse experts working on units dedicated to SCD treatment and management in the U.S. and Jamaica, and directors of community based organizations dedicated to education and assistance of those living with SCD. The participants have had greater than five years of experience in their current roles. Participants were not recruited from one particular institution; instead they were recruited from various institutions for their role in SCD treatment and management. Convenience sampling and "snowballing" aided in

recruiting participants. Because the focus of the study is on the education needs of adults with SCD it was important to include providers who had significant experience caring for this patient population, making it more likely that their input is accurate and reflective of the actual needs.

In addition, the researcher sought out input from two community based organizations whose mission statements are directly related to living with SCD and supporting families. This study includes a response from a director of a CBO dedicated to working with those living with SCD in the community. For the purpose of this article, the term "expert" will be used to refer to any of the participants. IRB approval was obtained prior to recruiting participants. The questionnaire was distributed with a disclosure statement attached. All parties voluntarily agreed to participate and no personal identifiable information was recorded or used at any time.

Data Collection and Analysis

Data collection took place in the summer of 2015. The questionnaire was distributed to the participants via email, and completed in whatever method they chose. Some participants chose to participate via phone interview and responses were transcribed verbatim at the time of the phone interview. This pilot study includes responses from both the United States and Jamaica.

An email correspondence was sent to RNs and APRNs working with adults living with SCD (nurse experts) asking them if they would like to participate in the pilot study. Executive Board Members of community based organizations were also asked to participate. Those who indicated that they would like to participate were sent another email with the disclosure letter and the questionnaire. APRN or RN respondents and CBO respondents completed similar questionnaires with questions focused either on the nursing perspective or the CBO perspective, respectively. They were asked to complete the questionnaire to the best of their ability and

return it to the researcher at their convenience. Requests by participants to complete the interview over the phone were honored and the researcher facilitated the phone interview by utilizing the questionnaire as a guide. Follow up questions were asked when clarification was necessary. All phone responses were transcribed immediately as the respondent spoke. As such, no audio-recording took place at any time. The questionnaires are located in appendices A and B. Excerpts from the responses are included in the discussion section of this paper using pseudonyms. This pilot study includes the responses of three participants.

Measure of Quality

Because of the qualitative nature of this pilot study, data saturation is the marker for sufficient data collection. This is identified by the researchers when "nothing new is emerging from the interviews" (Lobiondo-Wood and Haber, 2014, p.101). In this pilot study, many of the responses from the first two participants revealed common themes, however, data from a third interview was still incorporated to assure completeness.

RESULTS

Discussion

Nurse Healing is an advanced practice nurse working in an outpatient hematology clinic managing approximately 200 adults living with SCD. She sees her clients during pain crises, when refilling prescriptions, and of course, during well visits. In addition, she consults with other providers on an inpatient basis in the ICU, labor and delivery, and pain management. *Nurse Dedication* is the head nurse of a sickle cell clinic who also has a sibling with SCD. She has been the head nurse of that clinic for fifteen years. *Hope* is the executive director of a community based organization dedicated to promoting welfare for those living with SCD, and

has served in this role for six years. Furthermore, *Hope* is a carrier of sickle cell trait, and has family members with SCD.

"Most do not have the education they need to thrive with SCD" – Nurse Healing

Central to the purpose of this paper is the idea that there are limited publications that identify just what is required for the effective education of those living with SCD. *Hope, Nurse Healing,* and *Nurse Dedication* all agree that adults living with SCD are not well educated about their disease, and the quality of such education is variable. *Nurse Healing* explains that many providers are not educated themselves about SCD and its implications, making effective teaching impossible. In nursing school, *Nurse Dedication* was never taught specifically about educational needs of patients living with SCD.

"Some patients have difficulty understanding the information" –Nurse Dedication

Across the board, stigma was identified as a huge barrier to effective educational intervention. *Nurse Healing* explains: "People don't understand. When they see a low-income young black man seeking narcotic pain medicine in an emergency room, they are more apt to treat him like a drug seeker. There are no outward signs of a sickle cell pain crisis." *Hope* explains that those using medications to manage pain "may not be able to attend to learning or retain what they have heard." It is important for providers to realize opiate pain medication does diminish learning capacity.

To further compromise the possibility of effective patient teaching, microvascular strokes that have occurred throughout childhood often leave adults with SCD at a decreased ability to receive new, complex information. Unfortunately, many services available to those with

learning disability are no longer accessible once a person reaches the age of eighteen. *Nurse Dedication* identifies misunderstanding as a barrier to successful patient education and parents do not pass on vital information to their children regarding the diagnosis as the child grows up.

Following from the stigma and misunderstanding, both *Hope* and *Nurse Healing* identify provider mistrust as a barrier to patient education. *Hope* uses words such as "isolated" and "ostracized" to describe what it feels like for those living with SCD. She explains that some feel that they are being blamed themselves for experiencing pain. She says this is a "huge barrier" to educating victims of SCD. *Nurse Healing* concurs, "Patient/provider mistrust or misunderstanding absolutely seem to be barriers to patient education." She offers that there is a huge provider trust issue, which can be mitigated when the patient consistently sees the same person for care management.

"Meet the individuals where they are" – Hope

All of the respondents offered suggestions in improving the educational interaction between educator and individual with SCD. Visual materials, such as videos, brochures, skits, and graphics were identified by all respondents as effective methods. *Hope* offers that programs and events that engage people are most effective, such as bowling nights and exercise programs that also provide teaching related to the disease. Many community based organizations provide education at their events including walk-a-thons, health fairs, and family outings. *Hope* also says that social media can be an effective means of communication between individual with SCD and a support person.

To help make the complexities of SCD more understandable to the layperson, *Nurse Healing* advocates for the use of analogies. Along a similar line of thinking, *Hope* says that

technical language needs to be abolished when speaking to patients to promote absorption and understanding. The use of medical jargon not only will inhibit comprehension, but can also affect the trust relationship between client and provider. Repetition, reinforcement, and ongoing education at each interaction with someone living with SCD are keys to promoting acquisition of knowledge about SCD according to all of the experts interviewed.

Each respondent also included suggestions regarding the most important aspects of SCD requiring intense education for optimal quality of life. Their suggestions have been compiled and are included in Figure 1.

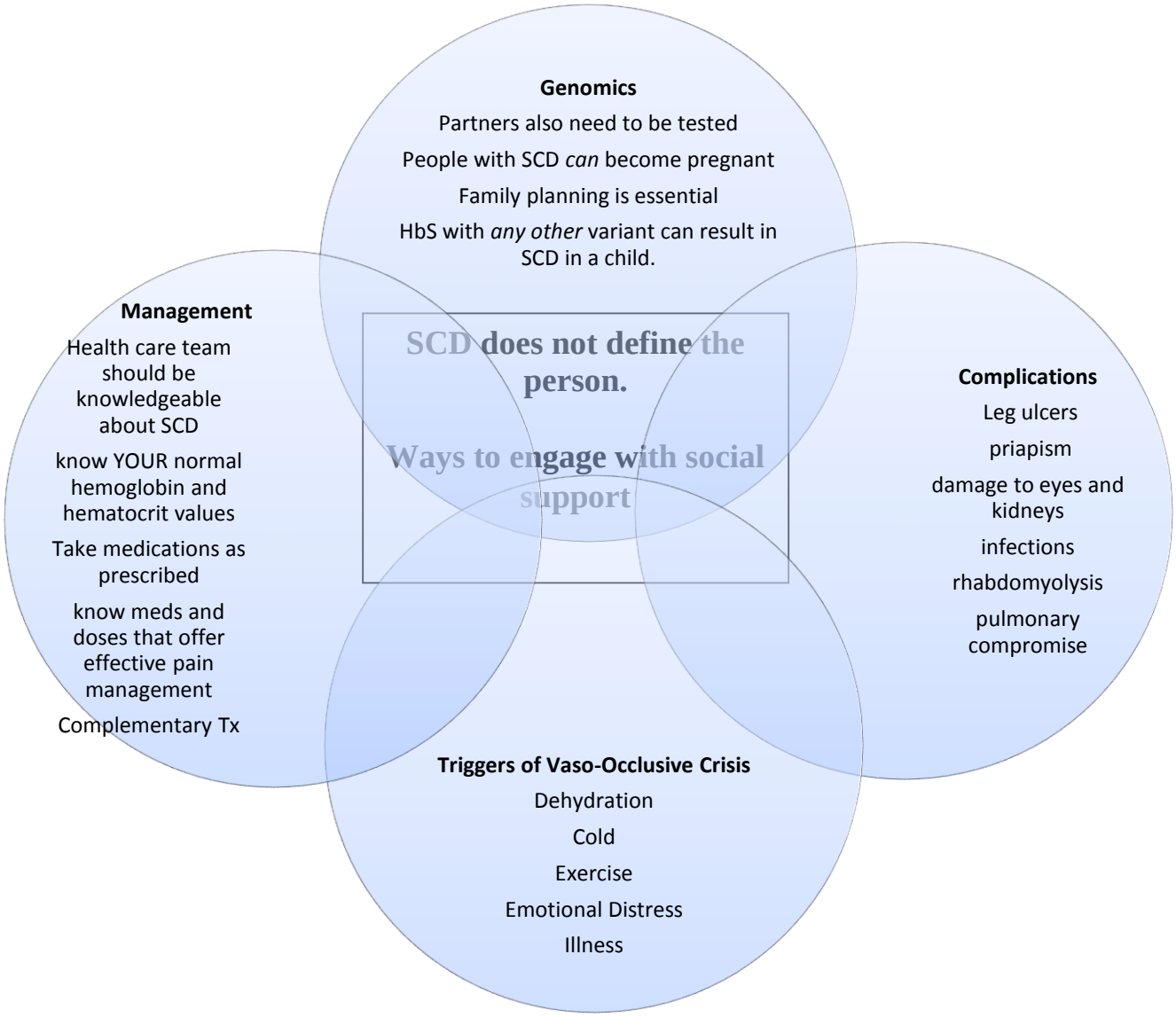

Figure 1

Expert identified components of quality SCD educational interventions

Implications for Clinical Practice

Patients must be made aware of the most important aspects of the disease. The results and discussion of this pilot study have presented what professionals in the field of SCD have identified as key points in education of those living with SCD. It is the hope of the authors that

the information provided in this pilot study will aid in the education of adults living with SCD for improved health outcomes and enhanced health related quality of life. Because of the severity of symptoms associated with sickle cell exacerbation, it is especially important to prevent these exacerbations. The information brought forth by this pilot study will guide the authors in developing a larger study with the aim of providing more evidence supporting components of high quality SCD educational materials.

Limitations

This pilot study is an unfunded BSN honors thesis. As such, sample size is relatively small compared to other similar studies. Additionally, this project was only afforded nine months to reach completion. Had the author had more time, a more complete analysis may have been made using a larger sample size. This pilot study and the expert responses and components of Figure 1 have the potential to provide a strong foundation for future studies investigating the educational needs of adults living with SCD, such as a quantitative analysis based upon the information gathered here.

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Appendix A

Provider Questionnaire

Demographic

1. What is your current role, and how long have you been in that role? Have you had any previous experience with sickle cell disease?

Interview

1. What are some of the most important things for an adult living with SCD to know about their condition?
2. Do you feel that patients are adequately educated about SCD, and have the knowledge they need to thrive with the condition? If not, what seems to be missing?
3. Were you taught specifically about educational needs of sickle cell disease patients? In nursing school or otherwise?
4. What are some teaching methods you've employed to help clients understand the process of SCD?
5. A. What barriers to educating adults with SCD have you encountered in your practice?

   B. Does patient/provider mistrust or misunderstanding seem to be a barrier to patient education?

6. A. Does there seem to be a need for teaching about genetics and how the disease is inherited?

   B. How about with respect to family planning?

7. Are patients educated about complementary and alternative therapies if they are thought to be effective?

Appendix B

CBO Questionnaire

Demographic

1. What is your role with GBSCDA and how long have you been in that role? Do you have previous experience with sickle cell disease, in another capacity?

Interview

1. What are some of the most important things for an adult living with sickle cell disease to know about their condition?
2. Do you feel that those with the condition are adequately educated about sickle cell disease, and have the knowledge they need to thrive with the condition? If not, what seems to be missing?
3. Does GBSCDA participate in educating those living with sickle cell disease? If so what kinds of teaching methods do you find effective? (i.e. guest lecturers, seminars, conferences, health fairs)
4. Did you receive any special training regarding sickle cell disease? Specifically, how best to educate those living with sickle cell disease about the condition?
5. A. Are you aware of any barriers that might make it difficult for someone living with sickle cell disease to understand their condition?

   B. Does mistrust or misunderstanding act as a barrier to teaching those with sickle cell disease about the condition?
6. A. Is it important for community members living with sickle cell disease to be educated about how the disease is inherited?

   B. How about counseling for family planning?

7. What are your thoughts about complementary therapies?