Identification of Knowledge Gaps in Adolescence with Sickle Cell Disease

Jennifer Hall

Rutgers School of Nursing

DNP Chair: Margaret Quinn, DNP, CPNP, CNE

DNP Team Member: Deepa Manwani, MD

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Abstract

Sickle cell disease (SCD) affects nearly 100,000 individuals in the United States, the majority being of African American and Hispanic/Latino descent. As recent as the 1970s, children with SCD often did not live past adolescence. Advancements in the identification and management of SCD have led to these children now living into adulthood. As children mature and parents become less involved in their care, they will need to assume responsibility for the management of their SCD as they transition from pediatric to adult health care. One of the barriers to a successful transition to adult care is insufficient disease knowledge and self-management skills. This project identified gaps in disease knowledge of adolescents aged 12-17 years via an online survey of SCD knowledge. The results of this survey were used to guide the development of an educational intervention to address the shortcomings in SCD knowledge with the intent of enhancing the transition process.

Keywords: sickle cell, adolescents, young adults, transition, knowledge, disease knowledge, education, chronic illness, chronic disease

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Introduction

Sickle cell disease (SCD) is an inherited blood disorder that causes a defect in the hemoglobin of red blood cells. This defect causes red blood cells to sickle – becoming rigid and misshapen like a crescent – when the body experiences specific stressors. These sickled cells can become trapped in smaller blood vessels, preventing proper blood flow throughout the body. This lack of blood flow not only leads to painful episodes in many different parts of the body, but can also lead to more severe acute and chronic complications when perfusion to the brain, lungs, kidneys, spleen, and liver are compromised (National Institute of Health, 2019).

Successful SCD management is essential in preventing painful episodes, infections, and long-term adverse outcomes that can result from organ damage. This success is, in part, due to adequate disease knowledge. An understanding of all aspects of SCD can provide a much-needed foundation for developing self-management knowledge and skills. Increasing this knowledge through education has the potential to not only improve patient self-management but possibly quality of life and health outcomes.

Providing education for caregivers is a process that begins when children are diagnosed with SCD, usually in infancy. As children mature, the responsibility of disease management shifts from the parent to the child, culminating in a transition from pediatric to adult health care. Therefore, it is essential to include them in conversations and education sessions, albeit in an age-appropriate manner. Providing disease knowledge is only one aspect of preparing adolescents for transition, but enhancing this knowledge can improve self-efficacy and set the stage for effective disease self-management, which is essential as they begin the transition process from pediatric to adult care (Centers for Disease Control and Prevention [CDC], 2018).

This project assessed the SCD disease knowledge of adolescents in specific domains (i.e., genetics, disease pathophysiology, symptoms/management, triggers/prevention, complications, and treatment) utilizing an approved questionnaire. The identification of specific knowledge gaps helped to facilitate the development of an age-appropriate, targeted educational intervention for adolescents with the goal of assisting them in building a solid foundation of disease knowledge during the early stages of transition planning. This education effort also has the potential to contribute to meaningful changes in health outcomes in the affected group, improving quality of life, life expectancy, and reducing overall healthcare costs.

Background/Significance

Sickle cell disease affects approximately 100,000 individuals in the United States (U.S.) most of whom are of African and Hispanic descent (CDC, 2017a). Until about the 1980s, the life expectancy for those with SCD in the U.S. was dismal with most children living only to the teenage years (Platt, 2018). Early identification through newborn screenings, immunizations, transcranial doppler screenings, blood transfusions, and the introduction of new medications have improved the life expectancy of children with SCD (Platt, 2018). About 90% of children are living into middle adulthood with a median survival between 58-66 years of age, depending on the specific sickle cell genotype (American Society of Hematology 2016, Elmariah et al., 2014). Despite living into adulthood, these patients still have a shorter life expectancy of 20-30 years than the general population without SCD (Lanzkron, Carroll, & Haywood, 2013).

This successful increase in life expectancy does not come without additional problems. An examination of mortality trends between the years 1979-2005 by Lanzkron, Carroll, & Haywood (2013) showed that while the rate of yearly pediatric deaths from SCD decreased by 3% each year, the death rate among adults increased 1% annually. Without disease

knowledge and self-management skills, providers alone will not be able to rectify their patients' poor outcomes (i.e., living longer does not translate to living better), leading to hospitalizations, surgeries, blood transfusions, and other costly medical interventions in adults with SCD. The annual healthcare cost of approximately $2.98 billion is, in part due, to SCD and its complications becoming more complex as people age (Huo et al., 2018; CDC, 2017b). Management of a chronic, complex disease can seem a daunting task to many and failure to manage SCD can lead to profound morbidity and mortality, but early and frequent education sets the stage to avoid these complications as disease knowledge can lead to better disease management. In short, education is a crucial component of preventative care.

The time of transition in adolescents with SCD is often a time of increased health care utilization and costs as well as an increase in morbidity and mortality, highlighting a need for improved efforts in this age group (Blinder et al., 2013; Quinn, Rogers, McCavit, & Buchanan, 2010). Adolescence is an ideal time to intervene to improve adult health outcomes and engaging adolescent patients in their care, educating them about their disease, and showing them how they can play a role in staying healthy has shown to be worthwhile (Zhou, Roberts, Dhaliwal, & Della, 2016; Gray, Schaefer, Resmini-Rawlinson, & Wagoner, 2018). As children with SCD live into adulthood, we must determine the most effective way to educate these pediatric patients earlier in life to affect adult outcomes. As the transition of care in SCD shifts from parents and pediatric providers to self-reliance and adult care providers, it has been determined by adolescents themselves to be a precarious time fraught with gaps in knowledge (Calhoun, Luo, Baumann, James, & King, 2018). As a result, new interventions must be identified and incorporated into the standard of care. Finding better ways improve disease knowledge and self-efficacy as it relates to disease management, may keep adolescents with SCD healthier, improve

quality of life, drive down healthcare costs, and potentially have implications for adolescents with other chronic diseases. In addition to the amelioration of poor long-term outcomes, these education efforts will likely also have near-term benefits as these adolescents become able to implement better disease self-management into their daily lives. This may lead to decreases in healthcare costs, days of missed school or work, financial and social-emotional burden, and decreased productivity in both adolescents and caregivers. In an increasingly outcomes-driven healthcare system, educational interventions sync with health policy, federal legislation, and goal statements, like the Triple Aim and Healthy People 2020 (Agency for Healthcare Research and Quality, 2017).

Needs Assessment

Globally, SCD occurs in approximately 300,000 births each year with most of those births occurring in Africa, the Middle East, and South Asia (American Society of Hematology, 2016). Unfortunately, most children in these regions lack the most basic healthcare. About half of the children with SCD in Africa will not live past the age of 5 years old and 20% of children with SCD in India will not live past the age of 3 years (American Society of Hematology, 2016). While an assessment of the need for education is an essential step in addressing disease self-management in adolescents, it is difficult to apply this needs assessment to many areas of the globe with underdeveloped healthcare and lack of resources where many children are succumbing to complications of SCD before they even reach adolescence. The focus for many of these countries is disease awareness, screening, and access to care (Biswas, 2013). Improvements in these healthcare systems will inevitably lead to a need for more education, not only in adolescents but the SCD population as a whole, for effective disease management. In developed countries with more robust health care systems, children with SCD are living into

adulthood necessitating a needs assessment of facilitators and barriers to improving disease knowledge with the goal of effective disease self-management (American Society of Hematology, 2016; Perry et al., 2017).

Sickle cell disease affects approximately 100,000 individuals in the United States most of whom are of African and Hispanic descent, and this number will continue to rise with birth rates of 1 in 365 for African Americans and 1 in 16,300 for Hispanic-American (CDC, 2017a). With an estimated individual lifetime cost for disease-related healthcare over $1 million, SCD warrants attention (American Society of Hematology, 2016).

Between 2000-2008, there were 1,911 infants born with SCD in New York State with birth rates much higher than the national average—1 in 230 infants born to African Americans and 1 in 2,583 infants born to Hispanic-Americans (Wang et al., 2013). During the years 2004-2008, there were 8,374 people with SCD living in New York, 40% of whom were under 21 years of age, 46% of whom were between the ages of 21 and 50 years old, and 14% of whom were older than 51 years (CDC, n.d.). Additionally, almost 50% of people with SCD aged 0 to 50 years had at least one emergency room visit, and 30% of all people with SCD had at least one hospital admission in 2008 (CDC, n.d.).

Of the 1,911 infants born in New York State with SCD, approximately 69% of these births occurred in New York City (Wang et al., 2013). These SCD statistics for New York City were not further dissected to identify births in each of the five boroughs, and thus specific data pertaining only to the Bronx was not available. The Bronx is a densely populated borough with a large cohort of individuals of African American and Hispanic-American descent. The racial/ethnic demographics of the Bronx as compared to New York City are as follows: New York City has 24.3% African Americans and 29.1% Hispanic-Americans while the Bronx has

43.7% African Americans and 56.2% Hispanic-Americans (United States Census Bureau, 2018). This racial/ethnic breakdown may increase the likelihood of many more individuals being born with SCD in the Bronx.

The exact number of children with sickle cell in the Bronx is unknown. Within the Bronx community, the outpatient hematology clinic where the project was carried out, serves approximately 700 patients with sickle cell disease, of which approximately 300 are adolescents who are in the transition period. The transition period should begin around the age of 12 years with preliminary discussions about the concept and policies surrounding transition and continue until the adolescent is established with an adult health care provider, ideally by a maximum age of 26 years (Got Transition, 2019). During this time, adolescents should be prepared for all aspects of transition planning and a large component of that planning is providing education.

In the setting of the outpatient hematology clinic, parental education begins at the first clinic visit and continues until the day of transition to adult care. During each visit, age-appropriate sickle cell education is provided to children and adolescents as well. With over 700 pediatric patients with sickle cell disease and a limited number of providers in a time-limited encounter, it can prove challenging to provide all the necessary education to manage a complex, chronic disease. Different avenues for providing education are needed to enhance learning and promote disease knowledge and self-management skills. Assessing the current disease knowledge of patients with SCD and developing an appropriate educational intervention will do just that. This approach coincides with the hospital's mission— “to heal, to teach, to discover, and to advance the health of the communities we serve” (Montefiore, 2019, para. 2).

Purpose

Lack of disease-specific knowledge in pediatric patients with SCD can lead to poor

disease management and subsequently poor outcomes. The purpose of this project was to assess the current disease knowledge of adolescents with SCD aged 12-17 years and identify gaps in specific domains. This information was then used to assist with the development of an age-appropriate educational intervention which targets the identified common knowledge gaps.

This project focused on answering the following question: In adolescents with sickle cell disease aged 12-17 years, will identification of disease-specific knowledge gaps and development of an age-appropriate educational intervention enhance the transition process from pediatric to adult health care services?

Aims and Objectives

The primary aim of this DNP project was to assess current disease-specific knowledge of adolescents aged 12-17 years living with SCD. The objective used to meet this aim included using an online anonymous survey which identified existing knowledge gaps within this population. The secondary aim of this project was to use this information to develop an educational intervention that directly targeted these gaps. Objectives included analyzing the data from the online survey, exploring age appropriate learning interventions that are fun and engaging, and creating a tool to be presented to the Division of Pediatric Hematology/Oncology for use with these patients to enhance the transition process.

Review of Literature

A literature review was conducted to find evidence of sickle cell disease knowledge, particularly as it pertains to adolescents with SCD during the transition from a pediatric to adult health care model. A search was conducted with the following databases: PubMed, CINAHL, Science Direct, OVID/Medline, and the Cochrane Library.

Initially, search terms focused on sickle cell disease but later came to include other chronic illness/disease due to the limited amount of transition-related issues pertaining specifically to sickle cell disease. Expanding the search allowed for a better overall representation of transition issues. The searches utilized the key terms sickle cell, adolescents, young adults, transition, knowledge or disease knowledge, education, chronic illness, and chronic disease in different combinations to collect literature between the years 2010-2019. The searches yielded 85 articles.

Inclusion criteria consisted of articles (1) in English, (2) focused on adolescents or young adults, (3) during the transition from pediatric to adult care, and (4) who have sickle cell disease or another chronic disease process. Articles were excluded if they were duplicated, could not be obtained electronically, or if they did not include relevant information pertaining to disease knowledge or self-management skills. The remaining 14 articles are included in this literature review (see Appendix A).

Transition

The life expectancy of children with SCD has increased dramatically with improvements in screenings, preventative care, medications, and other treatments leading to more than 90% of these children living into adulthood (Platt, 2018; American Society of Hematology 2016). These children with SCD who are now living into adulthood now need a more comprehensive understanding of their disease and how to manage it as they transition to adult care.

Transition describes a purposeful movement from pediatric care to adult-focused health care with intention "to maximize lifelong functioning and potential through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood" (American Academy of Pediatrics, American

Academy of Family Physicians American College of Physicians-American Society of Internal Medicine, 2002, p. 1304). Transition is not an event but rather a complex process that involves several stages ranging from the discovery of the concept of transition to preparation and planning to eventual transfer to adult care with feedback about the process (Got Transition, 2019). An important stage of the transition process is the preparation for eventual transfer to adult care and for this, adolescents need to have adequate knowledge and an understanding of their disease. While this knowledge is not the only facilitator of a successful transition, it is an essential component needed to establish the skills necessary to assume responsibility for their health care needs.

Disease Knowledge

Sickle cell disease is a complex, chronic disease with a myriad of acute and chronic complications. Having a solid foundation of disease knowledge can assist in managing these complications, but many adolescents with SCD lack this basic knowledge. Several studies highlight a concerning lack of knowledge among adolescents: 37% could not identify their SCD phenotype, only 38% knew their baseline hemoglobin, only 50% provided details regarding their medical history, and 54% reported difficulty discussing ways in which sickle cell anemia differs from other types of anemia (Williams et al., 2015; Speller-Brown, Varty, Thaniel, & Jacobs, 2019); Mennito, Hletko, Ebeling, Amann, & Roberts, 2014). Adolescents with SCD also lack knowledge regarding heredity, the importance of following a treatment plan, disease complications, the benefits of medication adherence, and the appropriate use of pain medications (Cecilio, Pereira, Pinto, & Torres, 2018; Poku, Caress, & Kirk, 2018). Additionally, integrative and systematic reviews of pediatric patients with chronic diseases (not specific to only SCD)

underscore a global lack of disease knowledge among not only patients but also adult providers, as well as insufficient self-management skills (Gray et al., 2018; Zhou et al., 2016).

The literature highlights not only the existence of knowledge gaps but also the necessity of improving disease knowledge and self-management skills. In five studies with structured interviews and focus groups, adolescents expressed the need for more SCD knowledge with regard to symptoms and management, medication, and preventative measures for pain triggers as a means to improve self-management skills and enhance readiness for a successful transition (Porter, Graff, Lopez, & Hankins, 2014; Porter, Wesley, Zhao, Rupff, & Hankins, 2017; Mulchan, Valenzuela, Crosby, & Diaz-Pow Sang, 2016; Melita, Diaz-Linhart, Kavanagh, & Sobota, 2019). Mulchan et al. (2016) also engaged SCD clinical experts in structured interviews and these experts discussed the importance of increasing knowledge about SCD complications to encourage adolescents to participate in good health maintenance behaviors. Young adults who have already transitioned to adult care highlighted the importance for adolescents to educate themselves about SCD, strive to become autonomous, and take responsibility for their SCD (Mennito et al., 2014; Porter et al., 2017). The perspectives of siblings and caregivers also echoed the importance of understanding and managing SCD. Siblings voiced concerns about the adolescents' self-sufficiency (e.g., taking medication, avoiding pain triggers) while caregivers expressed concern about medication adherence and their ability to communicate their medical histories to adult providers and adequately advocate for themselves (Porter et al., 2014).

In contrast with much of the literature, one study by Sabota et al. (2014) reported high levels of disease knowledge and self-management skills while adolescents in studies reviewed by Poku et al. (2018) demonstrated appropriate health-promoting behaviors, the avoidance of pain triggers, and the use of non-pharmacologic interventions for pain control. Speller-Brown et al.

(2019) also reported a relatively high degree of age-appropriate SCD knowledge among children, adolescents, young adults, and caregivers in general, but gaps existed in recognizing signs and symptoms of pain and the appropriate time to seek medical assistance. However, this same study also highlights a disconnect between adequate disease knowledge and disease management—they may have a good understanding of disease knowledge, but this did not translate to effective application of this knowledge in their daily living experiences. While a few studies highlight the strengths of adolescent patients with SCD, much of the literature emphasizes an overall knowledge deficit in several domains persists and requires attention.

Education

Education is an essential component of health care and providing sufficient SCD education in the early stages of adolescence has the potential to create a solid foundation of SCD knowledge. This new knowledge can encourage adolescents to take control of their disease and participate in making appropriate health care decisions. Without this disease knowledge, patients may not understand how their choices can affect their health outcomes.

There is limited literature on the effects of educational interventions and their impact on SCD knowledge. A Cochrane review by Asnani, Quimby, Bennett, and Francis (2016) examined 12 trials of educational interventions with patients with SCD and their caregivers. These trials consisted of cognitive and psycho-educational based interventions which involved diverse teaching methods such as written materials, audio recordings, videos, and computer programs. The overall results from these trials indicate that the educational interventions improved SCD knowledge. A recent study among adolescents and young adults in a co-location transitional model—a model in which transitioning adolescents and young adults are seen by both pediatric and adult providers from age 17-25 years—showed a significant improvement in

knowledge of SCD pain and retention of this knowledge from pediatric to adult care (Nolan et al., 2018). This study was also able to link this improvement in knowledge to a decrease in hospital visits and increased adherence in outpatient visits which provides some support for decreasing cost and healthcare utilization.

In summary, there is a paucity of disease knowledge among adolescents with SCD and the importance of ameliorating these gaps is evident. Educating adolescents about SCD has the potential to be one of the most effective strategies for increasing disease knowledge and will serve as a precursor to effective self-management skills and improved self-efficacy with the intention of positively affecting health outcomes.

Theoretical Model

The Knowledge-to-Action (KTA) model is a practical, organized approach to knowledge translation (see Appendix B). There are two interconnected processes within this model. In the first process, knowledge is created through inquiry and synthesis before it enters a second process, the action phase, in which a specific problem is identified, implemented, and evaluated through a series of steps (Graham et al., 2006). Although this process tends to proceed sequentially, it also allows for continued reevaluation and refinement, making it an excellent model for facilitating change.

For this DNP project, the knowledge created and synthesized related to the facilitators and barriers of the transition process from pediatric to adult health care. This information led to the identification of gaps in SCD knowledge as one of several problems during the transition process. Within the action phase, the assessment of specific domains of knowledge gaps (e.g., genetics, pathophysiology, treatments/medications, complications, triggers, prevention) was used to guide the development of a targeted educational intervention to increase disease-specific

knowledge and enhance transition readiness. While not a component of this project, the educational intervention can be implemented and evaluated to determine its effectiveness and the need for any revisions—at which point the cycle would begin again.

Methodology

This DNP project assessed the knowledge gaps in adolescents with SCD and subsequently an educational intervention was developed to address such gaps. The project used a descriptive quantitative approach using an online anonymous survey. After obtaining IRB approval at both the project site and at Rutgers University, the adolescent patients were recruited for the project via their parent (or legal guardian, going forward referred to as parent). Any interested adolescents then completed the online survey about SCD. The results were analyzed to determine the areas with inadequate knowledge and the project leader then developed an age-appropriate educational intervention to address the adolescents' needs. Identification of actual knowledge gaps provided an opportunity to create a tailored approach to educating adolescents with SCD.

Setting

Permission was granted for this DNP project to be conducted in an outpatient pediatric hematology clinic at a large metropolitan hospital in the Bronx, New York (see Appendix C). The racial demographics of the Bronx are 43.7% African American and 56.2% Hispanic/Latino (United States Census Bureau, 2018) and these two groups represent the races/ethnicities in which the majority of SCD is found. This broad representation of African Americans and Hispanics leaves this urban clinic providing medical services to approximately 700 pediatric patients with SCD until the age of 21 years.

Population

Participants for this DNP Project represented a convenience sample of adolescents with SCD age 12-17 years. There are approximately 245 adolescents registered in the outpatient clinic in this age bracket. Each of these adolescents was identified through the hospital's electronic health record and recruited for participation in this project. A review of birth dates showed that several of these patients were no longer eligible having recently reached their 18th birthday. Additionally, some patients on this list had also transferred their care to other medical centers thus eliminating them as potential recruits. The total number of adolescents eligible for recruitment was 178.

Recruitment

The participants in this project represented a convenience sample of adolescents with SCD who are currently receiving services in the outpatient hematology clinic. Convenience sampling provides the most appropriate approach for this DNP project as the educational intervention is only be meant to target this specific group of patients with SCD in this setting. The use of nonrandom sampling does not allow for broader generalizability though this is not the goal of this project. Inclusion criteria were adolescents between the ages of 12-17 years with a diagnosis of SCD. Exclusion criteria were the inability to read and speak English and lack of internet access, which is needed to complete the online survey.

Recruitment initially occurred via a letter mailed to the homes of eligible participants. This primary method of recruitment was the most effective means of targeting all eligible adolescents rather than targeting them by chance based on a regularly scheduled clinic visit within a short period of time. Since the participants are not legally adults, this letter was addressed to the parents of the adolescents (see Appendix D) with an invitation to join. The

recruitment letter described the purpose, details of the project, and how to contact the project leader. It also provided the active web address for the survey. If the parent felt this project was appropriate for their child, they were then able to discuss with their child her/his interest in participating. A second round of recruitment letters were mailed two weeks after the start of the survey to maximize participation.

In addition to mailing recruitment letters directly to the homes of eligible participants, project information flyers were also posted at the outpatient clinic check-in desk, the waiting room, and exam rooms to facilitate awareness of this project. This secondary means of recruitment was intended to target those who may not have received the mailed letter (e.g., lost in the mail, incorrect address) or those who may have received the letter but have since forgotten about the project. Recruitment for this survey was open for four weeks with the goal of a 20% response rate, equal to 36 adolescents.

Consent and Assent

The U.S. Department of Health and Human Services, through the Office for Human Research Protections, protects human subjects in research with special considerations for vulnerable populations such as children (Office for Human Research Protections, 2016). The project leader understands the need for protection of the participants and has completed the Human Research course through the CITI Training Program as required by Rutgers University and [redacted] (see Appendix F).

Consent was obtained from the parents and assent from the adolescents. There was no written consent or assent, rather both were provided via electronic format embedded in the online survey. Consent (see Appendix G) and assent (see Appendix H) were written in accordance with the Rutgers IRB and [redacted] guidelines and tailored to

this specific project. The consents and assents were written in lay language at approximately the 8th-grade and 4th-grade reading levels, respectively. Parents and adolescents were informed of their rights as they relate to the consent to participate in research. Consent was voluntary, informed, and confidential. Parents and adolescents who participated in the study provided consent/assent electronically in SurveyMonkey. If either the parent or the adolescent declined to participate in the study, the survey ended immediately. If the adolescent had already begun the survey and decided s/he no longer wished to participate, s/he could have immediately terminated the survey by selecting the EXIT button. For questions which had already been answered, participants were not be able to withdrawal their responses once they were submitted because the data was collected anonymously, making it very unlikely to link participants to their responses after submission.

Risks and Benefits

Participation in this project presented minimal risks for the adolescents. One possible risk was the psychological risk of feeling disappointed if s/he was unable to answer a question on the survey. This experience is likely similar to feelings s/he may have when unable to answer a question on a test at school. Another possible risk could have been an unwanted disclosure of disease status. If the adolescent completed the survey in a public forum, there could have been a chance that others viewed the survey and its content. The project leader attempted to minimize this risk by suggesting the adolescent complete the survey at home or in an otherwise private setting.

There were no immediate benefits to the participants. The participants' results guided the development of a SCD education tool, which may or may not provide benefits to them in the future in the form of improved SCD knowledge.

Participant Costs and Compensation

Costs for participation in this project were minimal. There was no monetary cost to participate; however, personal time was considered a cost for this project. The adolescents assumed the majority of the 'time cost' when they completed the online survey which, according to the data, took the participants an average of 15 minutes.

Although parents have supported their adolescents' in this project, the work was done by the adolescents in the form of completing the survey. Thus, the potential for compensation was given to the adolescents. When the adolescents completed the survey, they had the opportunity to enter a raffle for a $50 gift card. There was one $50 gift card available, and the winner was chosen randomly with the use of an online random name generator in which only the participants initials were used. The gift card was then mailed to the winner.

Project Interventions

Project implementation began soon after Rutgers University and [redacted] IRB approval. Eligible participants were recruited via a letter mailed to the home and consent/assent obtained as previously described. The web address allowed the adolescent to access and complete the survey via an online, cloud-based platform, SurveyMonkey.

The survey questions were derived from a Sickle Cell Disease Knowledge Assessment and Management Tool (see Appendix I) created by a group of healthcare providers at the hospital where the project was implemented. It is not validated tool, rather a survey of knowledge that has been used at the hospital to assist in knowledge assessment and transition planning. This tool was initially implemented with older adolescents (18-20 years) before their

transfer to an adult hematologist in the context of a research project that evaluated the role of a transition navigator in improving readiness to transition. The text is written at a 4th-grade level and was an appropriate tool for this population of younger adolescents aged 12-17 years.

In addition to the questions about SCD knowledge, eight demographic and disease background questions were added to the beginning of the survey for use in future statistical analyses. A free text box was also used to allow the participant to indicate any other topics related to SCD they perceive as relevant that may not have been addressed in the survey. Three questions pertaining to survey format were added at the end of the survey. The resultant 32-question revised version of the tool reflects these changes.

The revised version of the tool was directly transcribed to SurveyMonkey (see Appendix J). The opening pages of the survey contained a welcome statement with information about the survey, the electronic consent/assent, and instructions on how to take the survey. If either consent or assent were declined, the survey would have automatically ended. The questions were numbered throughout to allow the participant to know how many questions remain at any given time. There was only one disease specific question per page which required an answer. Unanswered questions did not permit the participant to move forward in the survey and they needed to select an answer before moving to the next page. Participants were not permitted to move backward in the survey to change an answer.

It is recognized that this survey may have prompted the participant to have additional questions about their SCD. For this reason, a short message at the end of the survey encouraged the participants to follow-up with their hematologist with any questions and concerns they may have. At the end of the survey, there was a web link which directed the participants to a different page where they were able to enter their names for a chance to win the gift card. The raffle entry

was independent of the survey and the participants answers were not linked to the raffle entry page.

Outcomes to be Measured

This project identified knowledge gaps in adolescents with SCD. The survey measured the participants SCD knowledge in six domains: genetics, disease pathophysiology, symptoms/management, triggers/prevention, complications, and treatment. The survey was scored in the same way a standardized academic test would be scored with one correct answer for each question and higher scores showing better disease knowledge than lower scores. The overall scores reflect a broader understanding of SCD and while important, it is the domain or specific questions with the lowest score that was the area of focus for education development. There was more than one domain that reflected a significant lack of disease knowledge. In this case, the project leader met with the principal investigator to determine the most relevant domain to address first and discuss the best possible approach to providing education to the adolescents.

The development of an educational intervention for the adolescents was informed by the results of the online survey. The intervention addressed the specific knowledge gaps with the use of an age-appropriate learning activity. The educational intervention has been designed and it will be presented to the Division of Pediatric Hematology/Oncology for implementation with the targeted group of adolescents.

Project Timeline

It was estimated this DNP project would take approximately 12 months to complete from inception to completion as described in the project timeline (see Appendix K). Delays in the IRB process were longer than anticipated and the project required 17 months for completion.

The project began in January 2019 and the final DNP project presentation marked its completion in April 2020.

The pre-design phase occurred over two months. During this time, the project leader had several meetings and phone/email discussions with the DNP team members to formulate a concrete idea for a successful project. After establishing the project, the PICOT question was defined.

It was anticipated that the design phase would last approximately eight months but instead required 12 months. This phase consisted of an in-depth review of the literature, exploration of theories and frameworks that can best describe this project, and the drafting of a complete project proposal with multiple revisions. A project was then presented to the DNP team members for approval to submit to both the project site IRB followed by an administrative review with the Rutgers IRB. The process of IRB submission and approval of the site IRB lasted for approximately 3 months with subsequent review with Rutgers IRB lasting approximately 2 months. While waiting for approval, the project leader constructed the online survey in SurveyMonkey and explored options for educational interventions.

Once the IRB at the project site and Rutgers IRB approved the project, the implementation phase was begun. During this time, the project leader accessed the electronic medical records to obtain the names and addresses of the parents of eligible adolescents and recruitment letters were printed and mailed on February 3, 2020 marking the start initiation of this project. The survey remained open for four weeks and the project implementation concluded on March 1, 2020.

The evaluation phase was the last phase of this project and occurred during the final two months as noted in the timeline. During this time, the data was analyzed, and knowledge gaps

were identified. The project leader then developed an age-appropriate educational intervention for adolescents with SCD in the form of a storyboard. Final manuscript revisions were made in anticipation for the final DNP project submission and DNP project presentation which were completed in April 2020.

Resources

The project required only a few resources for implementation. There were costs associated with some of the resources and were paid for by the project leader (see Appendix L). The Division of Pediatric Hematology/Oncology provided most of the resources in kind, as this project is directly assisting the division in elements of transition planning for the participants engaged in this project.

Instruments/supplies. A computer is needed for internet access, data collection, analysis and storage, and access to other files which pertain to the project. The project leader has a personal laptop that will be used for these purposes and will purchase a USB drive for the storage of all project related data. Printers and paper will be needed to print the parent recruitment letters and envelopes, labels, and stamps will be needed to mail the recruitment information. The Division of Pediatric Hematology/Oncology will provide these supplies.

Tools. The original Sickle Cell Disease Knowledge Assessment and Management Tool was used to measure young adolescents' knowledge of SCD and self-management to assist with transition planning. This tool was provided by Deepa Manwani, MD, principal investigator, and revised for this project. A SurveyMonkey account was needed for the online administration of the survey. The Division maintains an account with SurveyMonkey, and the project leader was granted access to this account.

Educational Intervention. An educational storyboard was developed via Storyboardthat.com. To build a fully customizable storyboard that can be printed without watermarks required a fee of $9.99. This was paid by the project leader. The storyboard will be printed in color by the Division of Pediatric Hematology/Oncology.

Incentives. The incentive for completing the survey was an entrance into a raffle for a $50 gift card. The project leader purchased this gift card.

Time. The time needed for the collection and analysis of data and the development of an educational intervention rested with the project leader. The project leader will perform these tasks unpaid. The other DNP team members have agreed to contribute their time in kind for project consultation and understand the terms of this commitment presented to them at the beginning of the project.

Storage. No physical storage space will be needed for this project. Electronic files will be kept on a USB drive in the possession of the project leader.

Data Analysis

SurveyMonkey and Microsoft Excel were used in the collection and management of data. SurveyMonkey collected and stored the results of the surveys and provided some general descriptive statistics in the form of measures of central tendency. This data was directly exported to a Microsoft Excel file. After the data was exported to Excel, it was inspected for any errors related to omission or other inaccuracies. The data included: number of participants completing the survey, demographic/background data (i.e., gender, age, race/ethnicity, sickle cell genotype, chronic transfusions/hydroxyurea, number of hospitalizations, perception of disease control), and scoring for each question (whether the answer was correct or incorrect).

Descriptive statistics were used to analyze the overall survey scores, the specific scores of individual questions in each domain, and the average scores of each domain. The specific statistics used were measures of central tendency, most importantly the mean. Identification of the mean allowed for identification of the knowledge domain with the lowest test scores.

Data Maintenance and Security

All efforts were made to keep the participants' survey responses confidential. This survey is anonymous, and it is highly unlikely the survey responses can be linked with a specific participant via demographic data. There was a separate link at the end of the survey which allowed participants to enter their name to win the $50 gift card which was being offered as an incentive. To further ensure confidentiality, this link to the raffle was not linked to the answers of the participants.

The survey data was stored in a password protected file on a USB drive and accessed on password protected computer. The SurveyMonkey account was also password protected. At the completion of this study, the USB drive with the survey results was securely stored at Rutgers University for six years, as required by the University, at which point it will be destroyed. The DNP team member at the project site has keep a copy of the data on a password protected computer at the [redacted]. In the future, this data may be used as a comparison with future survey results, and this is indicated in the consent form.

Results

Between February 3 and March 1, 2020, one hundred seventy-eight adolescents were recruited for this survey. Seventeen adolescents completed the survey for a completion rate of approximately 10%. All participants completed the full survey, meaning they did not exit the survey prematurely and answered all required questions. The demographic data of the

participants consisted of the following: eleven of the participants were female and six were male; Twelve were African-American, four were Hispanic/Latino, and one identified as other (African); Eight participants had hemoglobin SS disease, four had hemoglobin SC disease, and four did not know their sickle cell genotype.

This survey was scored as a test is scored because the questions pertaining to disease knowledge had correct answers. The overall mean score on the survey was 75% with a median score of 78%. There was a wide range of scores between 44% and 94%. The mode reflected two of the most common scores—94% (n=3) and 83% (n=3). Additionally, two other adolescents scored 89%. In four of the domains (genetics, pathophysiology, symptoms/management, triggers/prevention) the average score among all participants was above 80% with a range of 82%- 88%. The two domains with the lowest average scores were the domains of complications and treatment with scores of 49% and 55%, respectively. It was these two domains that were the target of patient education.

Discussion

The purpose of this project was to assess the current disease knowledge of adolescents with SCD and identify gaps in specific domains. Both overall knowledge and specific gaps are discussed here.

Overall Knowledge

With the overall disease knowledge represented by an average score of 75%, a median score of 78%, and a mode of 83% and 94%, it would appear these scores suggest an adequate overall knowledge base among adolescents in this clinic. As previously mentioned, this test was scored in the same way an academic test would be scored. If 70% is assumed to be an average score, as it is in most academic settings, then these results may be misleading since there were

still 41% of adolescents scoring below 70% with scores ranging from 44%-67%. A Pearson correlation was tested to ascertain any relationship between the age of the adolescent and the overall survey score, and no correlation was found ($r = 0.04$). It was thought that the younger adolescents may need more education about SCD, but this data highlights the need for ongoing education among all ages of adolescents with SCD.

Weakest Domains

The domains of disease complications and treatment were identified as having the weakest knowledge base as indicated by scores of 49% and 55%, respectively (Table 1). The average scores for each question in these domains can be found in Figure 1. As with the overall score, it was thought that the younger adolescents may have more knowledge gaps in the weakest domains than the older adolescents since each additional year represents more time living with a chronic disease. Additionally, the domains of complications and treatment may present more complex topics than the other domains and it was hypothesized that older adolescents may have a better understanding of these topics. A positive correlation may have supported these presumptions and may have indicated the need to intensify the targeted educational intervention in the younger adolescents. However, as with the overall scores, there was no correlation between age and average scores in the complication domain ($r = -0.04$). There was a weak negative correlation between age and scores in the treatment domain ($r = -0.38$), meaning the older the adolescent, the lower the score. This was, however, a very weak correlation that could have been influenced by many factors, including the outlier scores—the perfect score of 100% by the youngest adolescent as well as both of the oldest adolescents scoring 0%. Regardless of any weak or non-correlation, these results indicate the need for targeted education in these domains.

Free Text Answers

There was one free text question in the survey that asked the adolescents to identify the topic about SCD which they found most important. There was potential for this free text question to identify other areas of SCD that may not have been considered when developing the survey. The answer to this question did not yield any one overriding theme and most of the issues mirrored the domains reflected on the survey. One adolescent mentioned she would like to know more about issues related to self-management skills and understand how SCD could impact her in many different areas of life, "…I would also like to know things about how to deal with doing certain things outside of the medical obstacles of my sickle cell (school, going to college, traveling own your own, birth complications, etc.)." This is important information as it shows a desire to apply the knowledge she already possesses. This is the next important step with these adolescents who will need to build a foundation of knowledge and then learn to apply it prior to their transition to adult health care services. Another adolescent used this opportunity to reaffirm what she already knows about sickle cell disease and the ways in which applies that knowledge to her everyday life,

> If you have sickle cell disease you should make sure you know your body's limits. Take breaks when you need to, drink lots of water and take your vitamins. Make sure you tell your school nurse/gym teacher about it too so they can alert your parents if anything is wrong.

Finally, another adolescent used this opportunity to speak to difficulty of living with a chronic disease, "It's hard living with it." All these issues highlight not only a need for improving basic SCD knowledge but also the need to provide education on applying this knowledge to lived experiences as well as the much-needed psychological support for managing a chronic disease.

Limitations

There are a few limitations to this project. The major limitation was the low response rate. The success of this project was dependent on effective engagement with the targeted population. Sufficient recruitment and adolescent follow-through in taking the survey was essential to accumulate enough data to identify knowledge gaps. The insufficient response rate may not have provided enough data to adequately reflect true knowledge gaps.

Secondly, the survey was not based on a validated tool. The survey was developed by the providers in the clinic and modified by the project leader to create a short assessment of SCD knowledge. The survey consisted of only three questions in each of the six domains. It can be argued that this number is not enough questions to adequately represent knowledge in each domain and as a result the tool lacks content validity.

The survey was only offered in English. This many have served as an additional limitation. SCD is most common in those of African and Hispanic/Latino descent. Many of our patients are Spanish speaking and there are a smaller number of patients who are from Africa and English may serve as their second, and perhaps less proficient, language. These parents and/or adolescents may have opted out of the survey due to language barriers.

The lack of generalizability is the final limitation. This project was a quality improvement project designed to target education for the clinic's adolescent population. It was not expected that convenience sampling of one outpatient clinic would produce results that could be generalizable to a different population but the low response rate of this survey limits the ability to generalize about the larger population of adolescents aged 12-17 years.

Implications for clinical practice

The results of this project are in line with much of the literature. Many adolescents with

SCD are lacking basic knowledge about their disease and the skills needed to apply that knowledge to everyday life experiences. Education and anticipatory guidance are the foundation of most pediatric visits, whether in primary care or within the specialty services. Clinicians need to allow time to provide necessary SCD education to both parents and children through their lifespan, but this is not always possible within the time constraints of an average visit. For this reason, it would be important to identify additional points of contact during which teaching can occur as well as the best methods to deliver the education.

Additional points of contact that may be feasible for providing education would be times where patients are engaged for other treatments outside of their regularly scheduled visits such as a chronic transfusion appointment. Other patients also come to the day clinic for infusions of other IV medications for SCD. These transfusions and infusions provide time when the patient is waiting for the treatment to end—anywhere between 30 minutes to three hours. These periods of time can be used to engage patients with short periods of education. This education could be provided by the nurse, nurse educator, or the child life specialist if they have had training in teaching about SCD. If funding and resources are available, the clinic can consider a support group once or twice per month with time provided for education.

The best methods to deliver this education will vary by age group and change over time as technology changes. Basic information can still be provided in printed materials, but other media should be explored and possibly developed if funding permits. Printed materials can be lost or damaged and become increasingly less interesting with the ubiquitous use of electronic media. Many children and adolescents are now engaged in via electronic media with the use of mobile apps and the internet and this should be a consideration when providing education.

The method that will be used to deliver education targeting the gaps identified in this project will be a storyboard that will be printed and distributed to adolescents with SCD (see Appendix M). The storyboard uses pictures to describe common complications of SCD and the treatments or preventative care for each complication. The printed material will be made readily available in the outpatient clinic. It can be handed out by clinicians and other clinic staff and will also be on display for the patients and parents to take as needed. The project leader and the PI are also discussing the possibility of designing a free website with educational materials in the near future.

Implications for Health Care Policy

The purpose of this project was to assess and identify knowledge gaps with the intention of providing targeted education to adolescents in one clinic. The results from this project can be used as a model to guide education in other programs serving adolescents with SCD. Research can be done to determine the effectiveness of such education and what impact it may have on the transition planning process, the patients' successful transition to adult health care, and perhaps even the patients' long-term health outcomes. Having this data may also provide the much-needed support for the funding of SCD programs and research

Implications for Quality

Engaging children from a young age in discussions about their health has many benefits. Providing SCD education early and continuing throughout adolescents gives those with SCD disease the knowledge they need to understand their chronic condition and how to manage it. Encouraging these adolescents to take ownership of their SCD gives them a stake in their health and well-being. Arming them with the knowledge they need can may improve their self-confidence which can lead to better self-management skills. Effective disease management can

ultimately impact both the quality of their health and their quality of life—a process that can begin in pediatric care and carry them through adulthood.

Implications for Education

The success of any project is dependent on the buy-in of involved staff members. In this clinic the providers would need to be informed of the results of this survey and importance of providing effective education to adolescents with SCD. Additional staff members such as clinic nurses, inpatient nurses, nurse educators, and child life specialists are critical in the delivery of patient education and would also need to understand the significance of this project. Various methods will be used to disseminate the findings of this DNP project. At the project site, the results will be presented to the providers at the monthly Pediatric Hematology/Oncology Division meeting. Small group meetings with nurses will be held by the project leader in the hematology/oncology clinic and the inpatient unit and with child life specialists to discuss the results of this project and highlight the need for specific education. While the details of the project's results will not be directly disseminated to the participants and their parents, they will eventually be provided with the educational intervention which targets the gaps in SCD knowledge. This educational intervention will also be provided to patients of all ages or their parents if they are interested.

In the academic setting, the project leader presented the findings in a public forum at a scheduled DNP Poster Presentation Day at Rutgers University School of Nursing. This not only disseminated findings to stakeholders on the DNP team but also to others attending the poster presentations who may have had an interest in this topic. There is the possibility for future publication in a peer reviewed journal to further disseminate the results of this project.

Plans for Future Scholarship

This project identified SCD knowledge gaps in adolescents with the disease. The identification of these knowledge gaps resulted in an educational intervention to address these areas needing improvement. This was only the first step in preparing adolescents for transition and there are several avenues to ensure aspects related to this project are sustainable. To encourage the forward movement of this project, the developed educational tool can be implemented in the targeted population. Future projects could then evaluate the use of the effectiveness of the educational intervention within this population. Additional projects could focus on the evaluation of the Sickle Cell Disease Knowledge Assessment and Management Tool for reliability and validity. The cumulative effect of projects targeting patient education would further enhance the transition process.

Conclusion

Advancements in SCD have led to children living into well into adulthood. These children who often did not survive childhood now need to assume responsibility for their SCD as they transition to adult care. Lack of disease knowledge and self-management skills have been identified as one of the barriers to a successful transition of care. Providing adolescents with a strong foundation of disease knowledge early in the transition process can improve their understanding of SCD with eventual improvement in self-management skills. Ideally this may translate to better health outcomes marked by the increased length of life and improved quality of life.

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Table 1

Mean Scores by Age in Each SCD Domain

	Genetics	Pathophysiology	Triggers/ Prevention	Symptoms/ Management	Complications	Treatment
All ages	88%	88%	82%	86%	49%	55%
12 years	67%	100%	100%	67%	33%	100%
13 years	83%	83%	75%	83%	42%	25%
14 years	100%	89%	89%	78%	33%	78%
15 years	100%	100%	89%	100%	78%	67%
16 years	83%	83%	83%	92%	75%	50%
17 years	83%	83%	67%	84%	0%	50%

Note: Both the average scores of all the adolescents aged 12-17 years and the average scores for each age group are defined for each specific SCD domain.

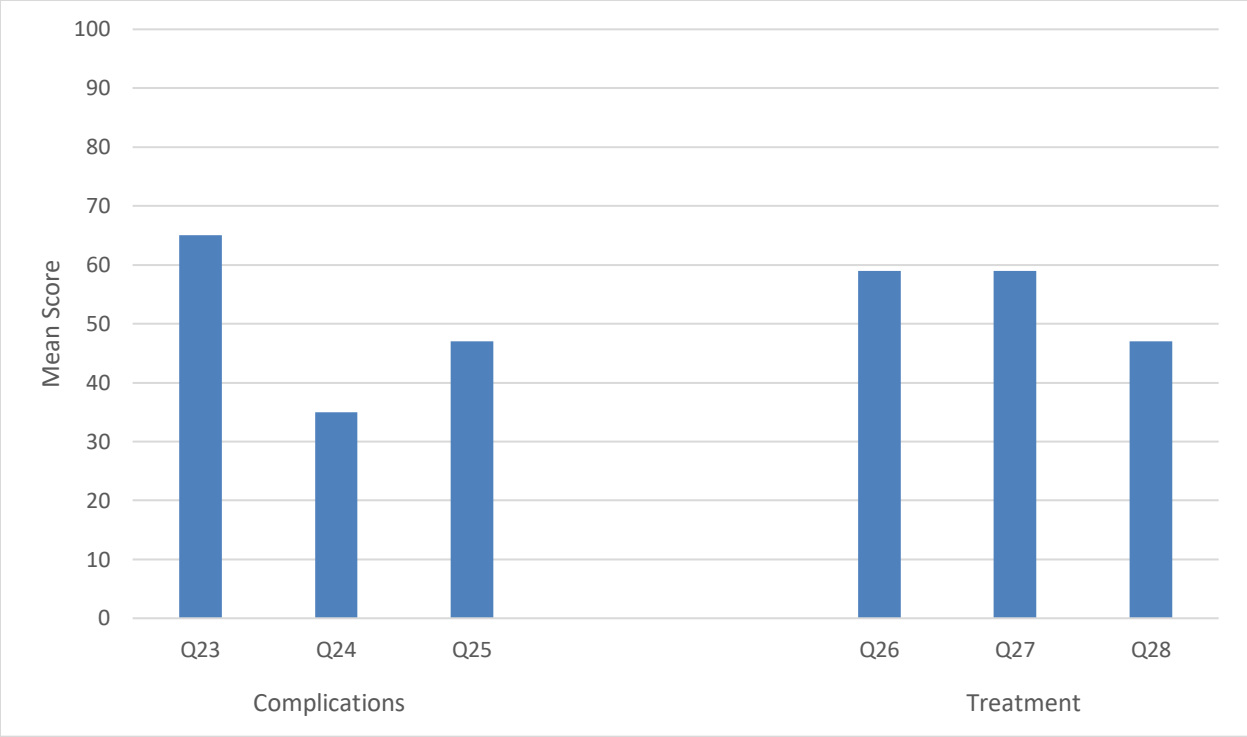


Figure 1. Chart representing the mean score of each question in the two lowest scoring domains.

Q23: Which of these is NOT a complication of sickle cell disease?

Q24: Avascular necrosis (AVN) is:

Q25: Which is a possible complication of chronic blood transfusions?

Q26: The only known cure for sickle cell disease is?

Q27: Hydroxyurea is a medication that can help sickle cell disease patient by:

Q28: What is the main treatment for preventing strokes?

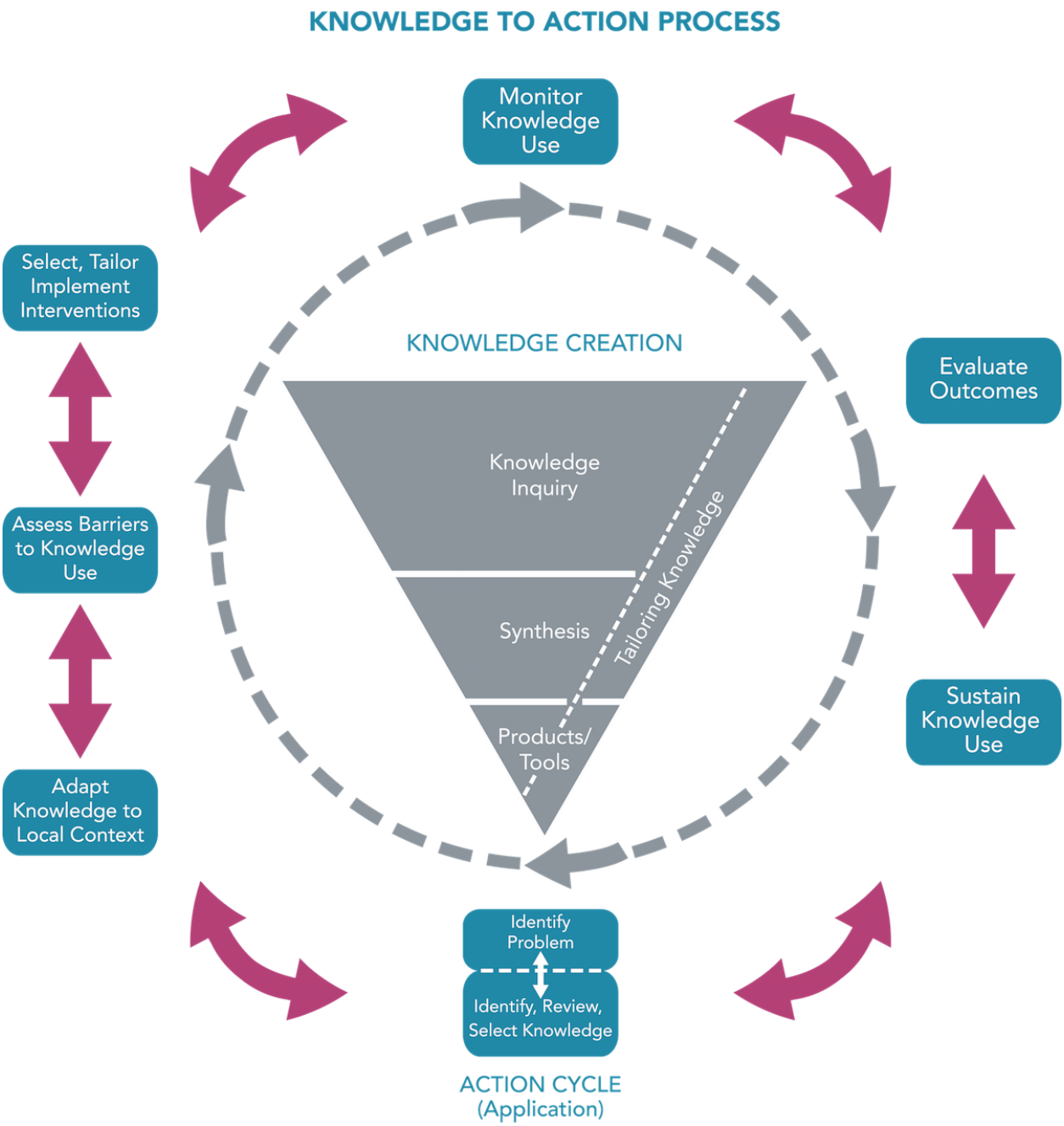
Appendix A
Table of Evidence

Article #	Author & Date	Evidence Type	Sample, Sample Size, Setting	Study Findings that help answer the EBP Question	Limitations	Evidence Level & Quality
1	Asnani et al., 2015	Systematic review	12 trials of 563 patients aged 6-35 years, mostly African Americans	Participants were randomly assigned to educational programs or a non-educational program. Interventions varied and lasted from 1 hour to weekly sessions for 8 weeks. Post-intervention assessments ranged from immediately after to 12 months. Educational programs showed improvement in knowledge of SCD. Four trials consisting of 160 participants directly assessed the SCD knowledge of pediatric patients age 6-18 years and showed a significant ($p<0.03$) improvement in knowledge after educational interventions.	Evidence ranged from low to moderate. The limited number of participants were too small to power the studies. High attrition rates may have contributed to small effect size. Lack of hand searching may have limited number of trials included in review.	I/B
2	Cecilio et al., 2018	Descriptive, exploratory qualitative study	17 AYA with SCD between 13-24 years from Outpatient's Clinic of [redacted] in Brazil	Each participant first completed a questionnaire for demographic data. They then had a 20-minute semi-structured interview to assess barriers to self-care and feelings associated with SCD. Five themes emerged: feelings, bullying/stigma, cognitive factors, medication compliance, and family issues. Within these themes, sub-themes emerged related to knowledge about medication, heredity, hydration.	Conducted in Brazil, may not be generalizable	III/C
3	Gray et al., 2017	Systematic Review	57 studies—24 quantitative, 25 qualitative, 8 mixed method Pediatric chronic illness population adolescents and young adults ≤25 years United States	Systematic review to identify barriers associated with transition from pediatric to adult care. Each disease has unique barriers but there are barriers common to all pediatric chronic disease related to relationships, beliefs/expectations, skills, transition knowledge, self-efficacy, access to care and insurance issues.	Only 12 studies focused on SCD 19 studies focused on knowledge barriers Only 2 focused on disease knowledge in SCD	II/B
4	Melita et al., 2019	Qualitative study	17 adolescents/young adults (AYA) between 13-18 years old with SCD and 15 caregivers of those AYA. Participants recruited from 2 pediatric SCD clinics in Boston, MA.	Five focus groups were used to identify facilitators and barriers of self-management to inform the creation of future problem-solving education intervention. The first half of the group involved AYA and caregivers but then the two populations split for the second half of group. Four major themes emerged: SCD management, transfer responsibility from caregiver to AYA, emotional/social support, and cultural expectations. Within this there were several subthemes—of importance, SCD knowledge.	Qualitative design in urban setting limits generalizability. Significant number of participants born outside the US.	III/B
5	Mennito et al., 2014	Non-experimental descriptive survey	3 adolescents and 5 young adults with SCD from a southern, rural pediatric practice	Surveys conducted to assess daily self-care issues, barriers to transition to adult care, satisfaction with transition experience, and input regarding creation of a transition program. Deficits found in SCD knowledge among other barriers and post-transition young adults provided feedback on needs for developing transition program (education for self and others about SCD, education on adult healthcare, assistance with disease management).	Small sample size- only one practice Rural population far from tertiary care center	III/C

Article #	Author & Date	Evidence Type	Sample, Sample Size, Setting	Study Findings that help answer the EBP Question	Limitations	Evidence Level & Quality
6	Mulchan et al., 2016	Descriptive, exploratory qualitative study	19 patients with SCD (8 adolescents and 6 young adults) from a pediatric and adult SCD clinic within a large southeastern medical center 10 clinical experts from various locations across the US	30-minute semi-structured interviews conducted with open-ended questions for both patients and clinical experts. Patients completed questionnaires assessing transition readiness. 10 themes emerged—one main theme being the importance of SCD knowledge as facilitator not only to transition but also to the development of self-management skills which is also a necessary component to transition.	Small convenience sample of mostly female participants and those who have had no stroke More adults than adolescents live below the FPL and group comparisons may not be accurate Those lost to transition cannot provide valuable information about their experience. Perspectives of family and caregivers not included	III/C
7	Nolan et al., 2018	Quasi-experimental pre/post-test design and descriptive correlational design	59 patients with SCD enrolled in a co-location clinic (intersecting pediatric/adult services) associated with [redacted] in TN	Two-part study: Education intervention: Patients in pediatric group (16-17 y/o) and adult group (18-19 y/o). Both given pre/posttest and 15-minute education about SCD pain. Significant improvement in knowledge in both groups. Analysis of health care utilization: pre/post-test scores used assess for association in improved knowledge and change in health care utilization between pre and post-transfer. The adult post-education scores were compared to post-transfer utilization. Study found association between higher post-education scores and lower hospitalization rates and increased health maintenance visits.	Small sample size limiting power. No comparison to those not in co-location clinic	II/C
8	Poku et al., 2018	Integrative narrative review	40 studies met inclusion criteria of: primary research, SCD patients between 12-19, patients' won reported experiences. US (24), UK (6), Jamaica (3), Sub-Saharan Africa (5), Middle East (2) Studies published between 1076-2016	All studies except 3 were of high to moderate quality. Nine broad themes emerged: knowledge and understanding, symptom experience, self-management, attitude to treatment, healthcare experiences, social relationships, difference and striving for normality, school experiences, emotional well-being/coping. In knowledge and understanding theme: patients have limited knowledge and understanding of heredity, SCD complications, nutrition, proper use of pain medications.	Only the first author assessed inclusion of potential studies. Lack of reporting of ample characteristics did not permit interpretation regarding demographic data. Lack of information about health care, culture, geography limit comparative studies.	II/A
9	Porter et al., 2014	Qualitative study	12 families consisting of: patient with SCD between 12-18 years, a sibling between 8-18 years closest in age to the patient with SCD with no chronic illness, and an adult primary caregiver. [redacted]	Focus groups conducted to explore the transition process from pediatric to adult care from the perspectives of the patient as well as the familial support system. Themes common among patients and family members: need for education about SCD knowledge and management skills, the ability of patients to care for themselves at the time of transfer, concerns about the knowledge and ability of adult providers to care for the adolescent/young adult.	Convenience sample may have only identified those likely to be engaged in patients' health care. Answers in focus groups may have been dominated by strong opinions or hesitancy to participate. Transition is a process with any stakeholders and health care providers were not included.	III/C
10	Porter et al., 2017	Qualitative and non-experimental descriptive	19 young adults (19-30 years old) with SCD who have transitioned to adult care; recruited from SCD community organizations, adult care providers, colleges, and SCD events in the mid-South region of the US	Qualitative component: 3 semi-structured focus groups discussed perceptions of transition, discussion about transition with others, and suggestions for improving transition process. Areas of SCD knowledge that were helpful in transition were identified. Quantitative component: Transitions Topics Checklist used to rate the importance of transition topics. Participants also asked to list the top 5 topics they felt were most important. Almost all topics scored as highly important for transition. Two of the top 5 most important topics are related to disease	Use of the SMART model is based on multiple stakeholders but here it included only the young adult perspective. Small sample size The majority were engaged in adult care and does not help in identifying barriers of those who are difficult to engage (e.g. those who refused and did not	III/C

				specific knowledge: understanding/taking medications and knowing SCD complications, understanding the impact of healthy/unhealthy behaviors	show) Some participants recalled experiences up to 12 years ago and some transition program may have changed.	
Article #	Author & Date	Evidence Type	Sample, Sample Size, Setting	Study Findings that help answer the EBP Question	Limitations	Evidence Level & Quality
11	Sobota et al., 2014	Non-experimental descriptive survey	33 young adults with SCD between 18-22 years from the [redacted] at [redacted]	Young adults engaged in a self-administered Sickle Cell Transition Intervention Program Skill Checklist assessing core areas for successful transition. The checklist assessed both knowledge skill sets and psychological components. Most patients had good medical knowledge of SCD but had gaps in knowledge in the areas of independent living and health benefits. Most were worried about transition, have sickle cell stress, or are concerned about being able to do the things they want.	Small, convenience sample Checklist is self-report which could lead to over-reporting positive results and not asked to demonstrate any skills.	III/C
12	Speller-Brown et al., 2019	Non-experimental descriptive	Large urban hospital serving 1700+ SCD patients birth-21 years 183 parents and/or children surveyed	Surveyed different age groups with age-appropriate surveys tailored to fit the disease knowledge and self-management skills for SCD. Several different ages targeted to assess across life span. Disease knowledge: across all age groups, there was a relatively high degree of disease knowledge to questions asked but this did not translate to adequate knowledge for disease management.	Small sample size, convenience sample at a single site. Limits generalizability Participants may have joined because they were confident in answering questions about SCD Used measurements that focused on self-report	III/B
13	Williams et al., 2015	Non-experimental descriptive survey	37 adolescents with SCD with mean age 14.9 years [redacted] onsite urban clinic and 4 off-site rural clinics	Surveyed adolescents to determine knowledge about transition process, preferred learning preferences, and understanding of SCD and self-efficacy. Only 43% were told about transition to adults and 21% recalled any related transition education. Patients prefer technology for learning such as online videos and smartphone apps in addition to being taught in clinic. Patients believed they were knowledgeable about SCD but wanted more information on long term effects of SCD on their organs and topics such as how to get health insurance, differences between pediatric and adult providers and between hematologist and other subspecialties. Most thought education should begin at 16 years.	Small sample size, with more surveys providers put more emphasis on education that researchers believed would lead to bias over time. Traditional didactic education was less effective than desired.	III/C
14	Zhou et al., 2016	Integrative Review	61 research articles from the US (31), UK (7), Canada (7), Netherlands (6) Nonexperimental quantitative (35), qualitative (15), mixed method (6), systematic review (5)	Focus of studies: chronic illness in general, disabilities, and diabetes. Six categories emerged: timing of transition, perceptions of transition, preparation for transition, patients' outcomes post-transition, barriers to transition, and facilitators of transition.	Search only referenced English documents which may have excluded other relevant studies. Half of the studies were non-experimental design that used self-report surveys.	III/A

Appendix B
Theoretical Model



CanChild. (n.d.) The Knowledge to Action Framework Process [Graham et al., 2006] [Digital image]. Retrieved from https://www.canchild.ca/en/research-in-practice/knowledge-translation-exchange

Appendix C
Site Letter of Cooperation

[redacted]

March 4, 2019

Re: Letter of Cooperation for [redacted]

Dear Jennifer Hall,

This letter confirms that I, as an authorized representative of [redacted] allow the Principal Investigator access to conduct study related activities at the listed site, as discussed with the Principal Investigator and briefly outlined below, and which may commence when the Principal Investigator provides evidence of IRB approval for the proposed project.

- [redacted]
- **Study Purpose:** The principle aim of this project is to identify knowledge gaps in patients age 12-15 years with sickle cell disease and develop and educational intervention to address these gaps.
- **Study Activities:** The principle investigator will obtain consent/assent and meet with participants and their guardian(s) to introduce and administer the knowledge assessment tool, collect and analyze data pertaining to the results, and develop an age appropriate education intervention to target identified gaps in knowledge.
- **Subject Enrollment:** Participants for this QI project will represent a convenience sample from patients followed in the outpatient hematology clinic. Inclusion criteria are: diagnosis of sickle cell disease (any genotype), between the ages of 12-15 years, and ability to read and speak English. The exact number of participants is yet to be determined.
- **Site Support**: [redacted] will permit physicians, nurse practitioners, and registered nurses from the outpatient clinic or inpatient unit to identify patients who may qualify for the QI project. The site will provide a quiet space for administration of the assessment tool.
- **Data Management:** Participants will be de-identified and any demographic information and data from assessments will password protected in all electronic formats (MS Word, MS Excel, SPSS, etc.). Paper assessments will be destroyed after all relevant information is entered into electronic documents, spreadsheets, or databases.
- **Anticipated End Date:** The QI project will end no later than May 1, 2020.

We understand that this site's participation will only take place during the study's active IRB approval period. All study related activities must cease if IRB approval expires or is suspended. I understand that any activities involving Personal Private Information or Protected Health Information may require compliance with HIPAA Laws and Rutgers Policy.

Our organization agrees to the terms and conditions stated above. If we have any concerns related to this project, we will contact the Principal Investigator. For concerns regarding IRB policy or human subject welfare, we may also contact the Rutgers IRB (see orra.rutgers.edu/hspp).

Regards,

[redacted]

[redacted]

Appendix D
Parent Recruitment Letter

[redacted]

Identification of Knowledge Gaps in Adolescents with Sickle Cell Disease

Hello,

My name is Jennifer Hall. I am a nurse on CHAM 9 and I am also a doctoral student at Rutgers School of Nursing. I am the Team Leader on a research study about sickle cell disease here at the [redacted] I would like to ask your permission to have your child participate in this study. Please allow me to give you a brief description of this study.

I would like adolescents to take an ***online anonymous*** survey about sickle cell disease. This survey will help me understand what they know about their disease and areas where they may need improvement. In the future, I may use the survey results to create an educational tool to help teach them about sickle cell disease.

To participate in this study your child must:

- Have sickle cell disease
- Be between the ages of 12-17 years
- Read and speak English
- Have access to the internet

The online survey should take less than 30 minutes. There is no immediate benefit to taking this survey. I will be offering an incentive for your child to participate. If s/he chooses to complete the survey, s/he will have an opportunity to enter a raffle for a $50 gift card. This raffle will have one winner and **NOT ALL PARTICIPANTS WILL RECEIVE A GIFT CARD**.

If you would like to have your child participate in this study, you should discuss it with her/him to see if s/he is also interested in participating. If you decide to take the survey, it can be accessed at:

www.surveymonkey.com/r/CHAM-SCD

I would be happy to speak with you and your child about the details of this study and answer any questions you may have. You can reach me by the email address or phone number listed below.

Thank you for your time.

Sincerely,

Jennifer Hall
[redacted]

Version 1.1 (9/22/19)

Appendix E
Clinic Recruitment Flyer

ADOLESCENTS NEEDED!

We want to know what **YOU** know about sickle cell disease!

This is a research study to find out what young adolescents know about sickle cell disease.

To participate you must:

- Have sickle cell disease
- Be between the ages of 12-17 years
- Read and speak English
- Have access to the internet

What do I have to do?

- You must first discuss this with your parents
- If you and your parent agree to participate, then you take an online survey
- It can usually be completed in less than 30 minutes

What's in it for me?

- Once you complete the survey, you can enter a raffle to win a $50 gift card!
- You can only complete the survey once!

CONTACT INFORMATION

Jennifer Hall

Version 1 (6/2/19)

Appendix F
CITI Training Certificate

Appendix G
Parental Consent

[redacted]

Parental Consent for Adolescent to Take Part in Research

Title of Project: Identification of Knowledge Gaps in Adolescents with Sickle Cell Disease
Principal Investigator: Deepa Manwani, MD
Student Researcher: Jennifer Hall, RN

Dear Parent,

The primary goal of this research project is to identify areas where adolescents may lack knowledge about sickle cell disease. We will use your child's results, in combination with the other children's results, to give us information about where to focus our teaching about sickle cell disease in young adolescents.

There are no immediate benefits to taking this survey. The results may lead to the future development of an educational tool that may or may not help to improve your child's understanding of sickle cell disease.

The risks in this study are very minimal. One possible risk is a psychological risk of a feeling of disappointment if s/he is unable to answer a question on the survey. This feeling may be similar to how s/he may feel if missing a question on a test at school. Another possible risk could be an unwanted disclosure of disease status if your child takes the survey in a public space. To minimize this risk, your child can take the survey at home.

This survey is anonymous, and your child's answers cannot be liked to her/him. If you do not want your child to take part in this study, you can choose "I DISAGREE" and you will automatically be exited from the survey. If you or your child have already started the survey and don't want to continue to participate in the study anymore, you can always click the EXIT button at the top of the screen. This will allow you to exit the survey but the questions you have already answered will be used in the research project.

If you have any questions about this research project, you can contact:

Jennifer Hall
[redacted]
Or
[redacted]

I allow my child to participate in this study. (Please select one answer.)

____ I agree

____ I disagree

Version 1.1 (9/30/19)

Appendix H
Adolescent Assent

[redacted]

Adolescent Assent to Take Part in Research

Title of Project: Identification of Knowledge Gaps in Adolescents with Sickle Cell Disease
Principal Investigator: Deepa Manwani, MD
Student Researcher: Jennifer Hall, RN

Hi!

We want to know what you know about sickle cell disease. There may be some things you don't know, and we want to find out what those things are. If we can do that, we can design fun ways for you to learn how to care for yourself.

Sometimes things happen to people in research studies that may hurt them or make them feel badly. These are called risks. The risk of this study is feeling down or disappointed if you don't know the answer to one of the questions on the survey. This may feel the same as not knowing the answer on a test at school.

If you take the survey in a public place, others may see what you are doing. If someone doesn't know you have sickle cell disease, they may find out if they see the survey. The best way to avoid this is to take the survey at home or in private.

This survey is anonymous. That means we won't be able to link your survey answers to you.

You don't have to be in this study if you don't want to. No one will get angry or upset if you don't want to be in the study. If you don't want to be in the study, you can choose "I DISAGREE" and you will exit the survey.

If you have already started the survey and don't want to continue, just click the EXIT button at the top of the screen. This will let you exit the survey but the questions you have already answered will be used in the research project.

If you have any questions about this research project, you can contact:

Jennifer Hall
[redacted]
Or
[redacted]

I understand what you told me and I want to take this survey. (Please select one answer.)

____ I agree

____ I disagree

Version 1.1 (9/30/19)

Appendix I

Sickle Cell Disease Knowledge Assessment and Management Tool (Original Version)

1. I have the following type of sickle cell disease:
    A) SS
    B) SC
    C) S-Beta thalassemia
    D) S trait

2. Sickle Cell Disease severity is variable. Are the symptoms from your Sickle Cell Disease
    A) poorly controlled
    B) not well controlled
    C) well controlled

3. How do you get sickle cell disease?
    A) from a virus
    B) from the water
    C) it is inherited from the parents
    D) from the hospital

4. I have the risk of having a baby with sickle cell disease if my partner has:
    A) SS
    B) SC
    C) S trait
    D) All of the above

5. Normal red blood cells are shaped like _____ and are flexible. Sickle shaped are shaped like a _____ and are rigid, hard, and sticky and can block blood flow.
    A) square, orange
    B) paper, apple
    C) discs, banana
    D) square, grape

6. How many cups of water should a person with sickle cell disease drink every day?
    A) 1-2 cups
    B) 8-10 cups
    C) It does not matter how many
    D) 4-6 cups

7. A person with sickle cell disease can drink alcohol and caffeine containing drinks?
    A) True
    B) False

8. Can a person with sickle cell disease smoke?
    A) No
    B) Yes, as long as it is tolerated
    C) It depends on the person
    D) Maybe

9. What can a person with sickle cell do to prevent a pain episode?
    A) Drinking sodas with caffeine
    B) Drink one 8-ounce glass of water daily
    C) Dress appropriately for the weather
    D) Smoking
    E) Get 4 hours of sleep nightly
    F) Drink alcohol

10. Which of the following do **not** trigger a crisis or pain episode?
    A) cold weather
    B) very hot weather
    C) windy weather
    D) rest and relaxation
    E) emotional stress and anxiety

11. A sickle cell patient should **urgently** see a doctor any time there is:
    A) Severe pain
    B) Fever of 101 or above
    C) Trouble breathing
    D) Weakness or paralysis of part of a body
    E) Any of the above

12. Draw a line to match the terms below.

A)	Stroke	Hard rock deposits in the gall bladder	G)
B)	Avascular necrosis (AVN)	Temporary or permanent loss of blood supply to bone	H)
C)	Acute chest syndrome	Damage to the eye	I)
D)	Retinopathy	Damage to the brain	J)
E)	Osteomyelitis	Pneumonia	K)
F)	Gallstones	Infection in the bone	L)

13. Draw a line to match the terms below.

A)	Signs of a stroke	Fever, stomach pain, nausea, vomiting, dark color urine, or clay colored stool	E)
B)	Symptoms of acute chest syndrome	Fever, cough, chest pain, wheezing, or shortness of breath and pneumonia	F)
C)	Symptoms of osteomyelitis	Paralysis to part of body, changes in vision/speech, or possible seizure, very severe headache	G)
D)	Symptoms of gallstones	Fever, swelling, soreness and heat to area	H)

14. When having pain, taking someone else's prescribed pain medication is ok.
    A) True
    B) False

15. The only known cure for sickle cell disease is
    A) Bone Marrow Transplant
    B) Blood Transfusion
    C) Hydroxyurea
    D) Natural remedies

16. Hydroxyurea is a medication that can help sickle cell disease patients by
    A) Decreasing pain episodes
    B) Prolonging life
    C) Improving energy
    D) All of the above

17. How confident are you that you can manage your sickle cell disease when:
    (Select the number that best represents your confidence level)

	Totally confident	Somewhat confident	Confident	Less confident	Not at all
A) You are not feeling well	5	4	3	2	1
B) You are given new medicines to take.	5	4	3	2	1
C) The weather has turned bad.	5	4	3	2	1
D) You are traveling.	5	4	3	2	1
E) You haven't been having any symptoms.	5	4	3	2	1
F) You're stressed out.	5	4	3	2	1

18. Rate yourself on how often you do the following statements.
(Select the number that best represents your answer)

	Always	Often	Sometimes	Rarely	Never
A) I remember to take my daily medicine.	5	4	3	2	1
B) I keep a schedule of when I am supposed to take each of my pain medicines.	5	4	3	2	1
C) I pay attention to my pain level before and 1 hour after I take my pain medicine.	5	4	3	2	1
D) I try not to let my emotions get out of control.	5	4	3	2	1
E) I know which of my symptoms mean I should get help right away.	5	4	3	2	1
F) I avoid triggers of pain such as dehydration, alcohol, smoking, over exertion, extreme weather (heat/cold) exposure.	5	4	3	2	1

19. How much do you agree with the following statements?
(Select the number that best represents your answer)

	Strongly agree	Agree	Disagree	Strongly disagree
A) Taking medicine every day is too much trouble.	4	3	2	1
B) I can manage my sickle cell disease without a doctor.	4	3	2	1
C) The medicines I take do me a lot of good.	4	3	2	1
D) I can handle most of the problems that sickle cell disease causes.	4	3	2	1
E) People can live normal lives with sickle cell disease.	4	3	2	1
F) Taking sickle cell disease medicine in public is embarrassing.	4	3	2	1
G) I know a lot about my sickle cell disease.	4	3	2	1

20. In the past year, have you had any trouble making or keeping appointments in regard to your SCD? (0 = no 1 = yes)

21. Do you take your own sickle cell disease medication?
    1. Never
    2. Sometimes
    3. Frequently
    4. Always

23. Do you feel stressed about your sickle cell disease?
    1. Never
    2. Sometimes
    3. Frequently
    4. Always

24. Are you physically active despite your sickle cell disease?
    1. Never
    2. Sometimes
    3. Frequently
    4. Always

25. Do you believe that your sickle cell disease can be controlled so that you have less symptoms?
    1. Never
    2. Sometimes
    3. Frequently
    4. Always

26. Do you believe that the sickle cell medicines that you take daily can cause problems?
    1. Never
    2. Sometimes
    3. Frequently
    4. Always

27. Do you feel helpless with your sickle cell disease?
    1. Never
    2. Sometimes
    3. Frequently
    4. Always

Appendix J

SurveyMonkey SCD Knowledge Assessment Survey

What do YOU know about Sickle Cell Disease?

Welcome to the Sickle Cell Knowledge Assessment Survey

Parents and adolescents should read these introduction pages together.

My name is Jenn Hall and you may know me as a nurse on CHAM 9. I am also a student at Rutgers School of Nursing. I am the Project Leader on this research study about sickle cell disease here at the [redacted]

This survey will help us identify what you understand about your disease and areas where you may need improvement. In the future, I may use the results of this survey to create an educational activity that I hope will improve your understanding of sickle cell disease.

Thanks again for participating in this survey. Your responses areSUPER important to us!

What do YOU know about Sickle Cell Disease?

Survey Instructions

This survey has 32 questions. The first two questions just ask you and your parent if you agree to participate. There is no time limit for this survey, so take your time. Each question must be answered to move onto the next question. If you don't know the answer, give it your best guess. There are two questions that require you to type in your own answer.

Please answer the questions ON YOUR OWN. If you need help with reading, you can ask your parent for help but make sure YOU choose the answer.

If at ANY TIME you do not want to continue, click theEXIT button on the top right of the screen. This will allow you to exit the survey but the questions you have already answered will be saved.

If you exit the survey before completing it, you will not be able to take it again.

If you complete the ENTIRE SURVEY, you will be able to enter into a drawing for a $50 gift card.

- **You should only complete this survey ONE TIME.**
- **You can only enter the drawing ONE TIME.**
- **The number of correct answers does NOT IMPROVE YOUR CHANCES OF WINNING.**

- The winner of the gift card will be chosen RANDOMLY.
- There will be ONLY ONE WINNER of the gift card.

What do YOU know about Sickle Cell Disease?

Parent Consent

Dear Parent,

The primary goal of this research project is to identify areas where adolescents may lack knowledge about sickle cell disease. We will use your child's results, in combination with the other children's results, to give us information about where to focus our future teaching about sickle cell disease in young adolescents.

There are no immediate benefits of taking this survey. The results may lead to the future development of an educational activity that may or may not help to improve your child's understanding of sickle cell disease.

The risks in this study are very minimal. One possible risk is a psychological risk of a feeling disappointment if s/he is unable to answer a question on the survey. This feeling may be similar to how s/he may feel if missing a question on a test at school. Another possible risk could be an unwanted disclosure of disease status if your child takes the survey in a public space. To minimize this risk, your child can take the survey at home.

This survey is anonymous and your adolescent's answers cannot be linked to her/him. If you do not want your child to take part in this study, you can choose "I DISAGREE" and you will automatically be exited from the survey. If you or your child have already started the survey and don't want to continue to participate in the study anymore, you can always click the EXIT button at the top of the screen. This will allow you to exit the survey but the questions you have already answered will be used in the research project.

If you have any questions about this research project, you can contact:

Jennifer Hall

[redacted]

Or

[redacted]

* 1. I allow my child to participate in this study.

- ◯ I agree
- ◯ I disagree

What do YOU know about Sickle Cell Disease?

Adolescent Assent

We want to know what you know about sickle cell disease. There may be some things you don't know, and we want to find out what those things are. If we can do that, we can design fun ways for you to learn how to best care for yourself.

Sometimes things happen to people in research studies that may hurt them or make them feel bad. These are called risks. The risk of this study is feeling down or disappointed if you don't know the answer to one of the questions on the survey. This may feel the same as not knowing the answer on a test at school.

If you take the survey in a public place, others may see what you are doing. If someone doesn't know you have sickle cell disease, they may find out if they see the survey. The best way to avoid this is to take the survey at home or in private.

This survey is anonymous. That means we won't be able to link your survey answers to you.

You don't have to be in this study if you don't want to **No one will get angry or upset if you don't want to be in the study. If you don't want to be in the study, you can choose "I DISAGREE" and you will exit the survey.**

If you have already started the survey and don't want to continue, just click the EXIT button at the top of the screen. This will let you exit the survey but the questions you have already answered will be used in the research project.

If you have ANY QUESTIONS **about this research project, you can contact:**

Jennifer Hall

[redacted]

[redacted]

Or

[redacted]

* 2. I understand what you told me and I want to take this survey.

- ◯ I agree
- ◯ I disagree

What do YOU know about Sickle Cell Disease?

Tell us about YOU!

* 3. What is your gender?

- ◯ male
- ◯ female

* 4. I am:

- ◯ African American
- ◯ Hispanic/Latino
- ◯ Other (please specify)

* 5. How old are you?

- ◯ 12
- ◯ 13
- ◯ 14
- ◯ 15
- ◯ 16
- ◯ 17

* 6. I have the following type of sickle cell disease:

- ◯ SS
- ◯ SC
- ◯ S beta thalassemia
- ◯ S trait
- ◯ I don't know

* 7. Do you come to the hospital each month for a blood transfusion (chronic transfusion)?

- Yes
- No

* 8. Do you take hydroxyurea?

- Yes
- No

* 9. How many times were you admitted to the hospital in the past year?

- none
- 1-2 times
- 3-4 times
- 5-6 times
- 7 or more times

* 10. Sickle cell disease severity is variable. Right now, would you say your sickle cell symptoms are:

- Poorly controlled
- Fairly well controlled
- Well controlled
- Very well controlled
- I don't know

What do YOU know about Sickle Cell Disease?

Time to talk about Sickle Cell Disease...

* 11. Who can get sickle cell disease?

- African Americans
- Caucasians
- People from the Middle East
- Anyone

What do YOU know about Sickle Cell Disease?

* 12. How do you get sickle cell disease?

- ◯ From a virus
- ◯ From the water
- ◯ It is inherited from the parents
- ◯ From the hospital

What do YOU know about Sickle Cell Disease?

* 13. I have the risk of having a baby with sickle cell disease if my partner has:

- ◯ SS
- ◯ SC
- ◯ S trait
- ◯ All of the above

What do YOU know about Sickle Cell Disease?

* 14. Normal red blood cells are shaped like _____ and are flexible. Sickle cells are shaped like a _____ and are rigid, hard and sticky and can block blood flow.

- ◯ Square,orange
- ◯ Discs, apple
- ◯ Discs, banana
- ◯ Square, grape

What do YOU know about Sickle Cell Disease?

* 15. Sickled red blood cells CAN prevent blood from reaching your muscles and organs.

- ◯ True
- ◯ False

What do YOU know about Sickle Cell Disease?

* 16. If red blood cells, and therefore oxygen and nutrients, do not reach your muscles and organs they will become:

- ◯ Stronger
- ◯ Damaged
- ◯ Unchanged
- ◯ Increase in number

What do YOU know about Sickle Cell Disease?

* 17. Drinking alcohol and caffeine containing drinks can trigger a crisis or pain episode.

- ◯ True
- ◯ False

What do YOU know about Sickle Cell Disease?

* 18. Smoking can trigger a chest crisis or acute chest syndrome.

- ◯ True
- ◯ False

What do YOU know about Sickle Cell Disease?

* 19. What can a person with sickle cell do to prevent a pain episode?

- ◯ Avoid exposure to very windy or hot weather and dress warmly for cold weather
- ◯ Avoid emotional stress and anxiety
- ◯ Get adequate sleep and rest
- ◯ Drink lots of fluids
- ◯ All of the above

What do YOU know about Sickle Cell Disease?

* 20. A sickle cell patient should urgently see a doctor any time there is:

- ◯ Severe pain
- ◯ Fever 101 and above
- ◯ Trouble breathing
- ◯ Weakness or paralysis of any part of the body
- ◯ Any of the above

What do YOU know about Sickle Cell Disease?

* 21. If your pain is really bad and you run out of pain medicine, taking someone else's pain medication is ok.

- ◯ True
- ◯ False

What do YOU know about Sickle Cell Disease?

* 22. What are signs of a stroke?

- ◯ Stomach pain after eating fatty foods, nausea, vomiting
- ◯ Fever, cough, chest pain, difficulty breathing
- ◯ Paralysis to part of the body, changes in vision/speech, very severe headache
- ◯ Fever, swelling, soreness and heat to affected area

What do YOU know about Sickle Cell Disease?

* 23. Which of these is NOT a complication of sickle cell disease?

- ◯ Eye problems
- ◯ Stroke
- ◯ Acute chest syndrome
- ◯ Bone infection
- ◯ Constipation

What do YOU know about Sickle Cell Disease?

* 24. Avascular necrosis (AVN) is:

- Hard rock deposits in the gallbladder
- Damage to the bone
- Damage to the eye
- Damage to the brain
- Pneumonia
- Infection in the bone

What do YOU know about Sickle Cell Disease?

* 25. Which is a possible complication of chronic blood transfusions?

- Anemia
- Dehydration
- Iron overload
- Pain

What do YOU know about Sickle Cell Disease?

* 26. The only known cure for sickle cell disease is:

- Bone marrow transplant
- Blood transfusion
- Hydroxyurea
- Natural remedies

What do YOU know about Sickle Cell Disease?

* 27. Hydroxyurea is a medication that can help sickle cell disease patients by:

- ◯ Decreasing pain episodes
- ◯ Prolonging life
- ◯ Improving energy
- ◯ All of the above

What do YOU know about Sickle Cell Disease?

* 28. What is the main treatment for preventing strokes:

- ◯ Hydroxyurea
- ◯ Folic acid
- ◯ Transcranial doppler
- ◯ Monthly blood transfusions

What do YOU know about Sickle Cell Disease?

29. Please tell us the most important thing YOU want to know about sickle cell disease.

What do YOU know about Sickle Cell Disease?

Just a few questions about this survey...

* 30. Were the survey questions easy to understand?

- ◯ Yes
- ◯ No
- ◯ Not sure

* 31. Did you need someone to help read the questions to you?

- Yes
- No

* 32. How can we make this survey better?

What do YOU know about Sickle Cell Disease?

THANKS!

Thanks so much for filling out this survey!

This survey may have left you with some questions about Sickle Cell Disease. If you have any questions, you can follow up with your provider in the clinic.

If you have questions about this survey, you can contact me. Here's my contact information:

Jenn Hall
[redacted]

What do YOU know about Sickle Cell Disease?

Raffle Time!

Here's the link to enter into the raffle to win the $50 gift card. Don't forget--there will be only ONE winner.

Raffle Link!

SCD Raffle

Good Luck!

Please give me your name and address so I can mail you the gift card IF you are the winner.

Your name is required so I can include you in the raffle. If you want to pick up the card instead, you can contact me to make arrangements:

Jenn Hall

[redacted]

Remember, your answers are NOT linked to your name and address!

* 1. First & Last Name

2. Contact Information

Address	
City/Town	
State/Province	
ZIP/Postal Code	

Appendix K
DNP Project Timeline

Completion	Pre-Design	Design	Implementation	Evaluation
January 2019	Discussions with RSoN Program Director to develop and finalize DNP project			
February 2019	Meetings and discussions with RSoN Program Director and Director of Hematology at prospective project side to discussion proposed DNP project Development of PICO question Formalized DNP Team	Review of supporting literature Draft first part of project proposal: background and significance, needs assessment, problem statement, aims and objectives Proposal revisions		
March 2019		In depth review of literature and synthesis of information Explored theories and frameworks which would support project Draft second part of proposal: literature review and theoretical framework Proposal revisions		
April 2019		Meetings with site team member to discuss details of implementation Explored use of SurveyMonkey		

		Draft third part of proposal: methodology, data analysis, and evaluation plan Proposal revisions Project proposal presentation		
May 2019		Create template of survey in SurveyMonkey		
June 2019		Submit to site IRB		
August 2019		Consider different options for development of educational intervention		
October 2019		Site IRB approval Submit to Rutgers IRB		
December 2019		Rutgers IRB approval		Data analysis Development and distribution of educational intervention Manuscript revisions Final DNP project submission
January 2020			Gather necessary contact information for recruitment letters	

February 2020			Recruitment Implementation Data collection	
March 2020				Data analysis Complete final draft of paper Design poster
April 2020				Final Presentation Development of Storyboard
May 2020				Close out with both IRBs

Appendix L
Budget for Resources

Personnel			
Name	**Role in project**	**Time/Effort**	**Cost**
Deepa Manwani	Principal Investigator	10%	In kind
Jennifer Hall	Project Leader	75%	In kind
Margaret Quinn	DNP Team Member	15%	In kind
Non-personnel			
Item	**Quantity**	**Cost**	**Payor**
Paper	1 ream	n/a	In kind from Hematology/Oncology
Toner (ink)	1	n/a	In kind from Hematology/Oncology
Envelopes	356	n/a	Jennifer Hall
Stamps	356	n/a	In kind from Hematology/Oncology
SurveyMonkey Account	1	$37/month	In kind from Hematology/Oncology
Storyboard Account	1	$9.99	Jennifer Hall
USB drive	1	$5	Jennifer Hall
Gift card	1	$50	Jennifer Hall

Appendix M
SCD Storyboard