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Adherence to Self-Care Management of Sickle Cell Disease Among Caregivers

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Walden University
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Abstract

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MSc, Olabisi Onabanjo University, 2010

Final Diploma, University of Lagos, 2002

Dissertation Submitted in Partial Fulfillment

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Abstract

The self-care management of sickle cell disease (SCD) improves mortality rate; however, compliance with SCD self-care management remains a problem. The purpose of this study was to examine the knowledge and factors that influence compliance with SCD self-care management recommendations among caregivers of children with SCD. The health belief model was used as the theoretical foundation of this study, theorizing that caregivers' perceived susceptibility, severity, and benefits of SCD self-care management will influence compliance. The study used a quantitative research design. A cross-sectional survey was administered to 100 caregivers of children with SCD attending sickle cell clinics in Lagos, Nigeria using convenience sampling. Information was obtained from participants using a structured interviewer-administered questionnaire, and data were analyzed using descriptive statistics, bivariate correlations, and binary logistic regression techniques. Findings confirmed a high adherence rate but low knowledge of SCD self-care management among the caregivers of children with SCD. There was no significant correlation between knowledge of SCD self-care management and adherence. However, the findings from the multivariate analysis identified knowledge as a predictor of adherence and religiosity and total number of barriers as barriers to adherence. Parental health beliefs did not influence adherence to SCD self-care management. These findings have social change implications by guiding the work of health educators, health care providers, and public health practitioners to incorporate group counseling on SCD self-care management at every sickle cell clinic.

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Dedication

My dad always wanted one of his children to become a medical doctor. Long ago, I promised him that he may not get a child who is a medical doctor, but he will surely get a child who will be called a doctor. This dissertation is dedicated to my daddy as a way to tell him that I kept my promise. I also dedicate this dissertation to every sickle cell disorder patient out there. Remember, you can be whatever you want to be irrespective of your disorder.

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Table of Contents

List of Tables

Chapter 1: Introduction to the Study

Background of the Study

Sickle cell disease (SCD) represents a group of inherited blood disorders that affect the hemoglobin within the red blood cells (Centers for Disease Control and Prevention [CDC], n.d.). SCD is a genetic disease caused by inheriting two abnormal hemoglobin genes, one from each parent (Behrens & Cymet, 2000; Sergeant, 2013). The abnormality seen in the hemoglobin gene occurs due to mutation (Behrens & Cymet, 2000; Sergeant, 2013; Stuart & Nigel, 2004). This mutation in the hemoglobin gene, in the case of SCD, causes the production of red blood cells that are sickled or crescent shaped, sticky, and rigid, instead of the round shape, smooth and flexible features seen in red blood cells formed from normal hemoglobin (Hemoglobin A). The stickiness, inflexibility, and shape of the sickled red blood cells makes the cells clump together and block the blood vessels, inadvertently blocking the flow of blood and oxygen throughout the body (Behrens & Cymet, 2000; Sergeant, 2013; Stuart & Nigel, 2004). The red blood has a shorter life-span when compared to normal red blood cells, which causes them to collapse and break down easily (National Heart, Lung, and Blood Institute [NHLBI], 2012; Stuart & Nigel, 2004). These characteristics of the sickled red blood cell are the major cause of the complications seen in people with SCD (CDC, n.d.; Johns Hopkins University, n.d.; NHLBI, 2012; Stuart & Nigel, 2004).

SCD results from inheriting a homozygous sickle cell gene (Hemoglobin SS), or as a double heterozygote, which is a sickle cell gene in combination with an interacting gene (Hemoglobin S and Hemoglobin C or Hemoglobin S in combination with Beta

Thalassemia). In cases where only one abnormal hemoglobin gene is inherited, this is referred to as carrying the sickle cell trait. Persons carrying the sickle cell trait have one sickled hemoglobin S and one normal hemoglobin gene. Persons with the sickle cell trait are most times asymptomatic and normally have life expectancy equal to that of persons with normal hemoglobin genes (Behrens & Cymet, 2000; Sergeant, 2013). However, couples both carrying the sickle cell trait have a 1 in 4 chance of having a child with SCD.

SCD is the most common of the inherited hemoglobin disorders in the world, affecting millions of people globally (CDC, 2011; Modell & Darlison, 2008; Piel, Hay, Gupta, Weatherall, & Williams, 2013; World Health Organization [WHO], 2015). The exact prevalence of SCD is unknown, even in countries like the United States (Hassell, 2010). However, SCD has been reported to affect 100,000 people in the United States (Hassell, 2010). The ease of migration has led to the spread of the sickle cell trait worldwide. The prevalence of SCD is highest among people from, or descendants of people from, sub-Saharan Africa; South and Central America; the Caribbean; Middle Eastern Countries like Iran and Saudi Arabia; some regions in India; and Mediterranean countries such as Italy, Greece, and Turkey (CDC, 2011; Modell & Darlison, 2008). SCD is particularly predominant in Africa, with about 75% of SCD cases occurring in Africa (Akinyanju, n.d.; Modell & Darlison, 2008; Sickle Cell Foundation Nigeria, 2011). The predominance of SCD in Africa can be attributed to the high distribution and carriage of the sickle cell trait in Africa and because the sickle cell trait confers protection to severe malaria, which is endemic in most African countries, and malaria morbidity (Aidoo et al.,

2002; CDC, 2011; Gong et al., 2012; Williams et al., 2005). A mapping of the global distribution of the sickle cell gene shows a strong relationship between malaria endemicity and the prevalence of hemoglobin S genes (Piel et al., 2010). This makes the geographical distribution of SCD analogous to that of malaria.

In Africa, the distribution of the sickle cell trait and subsequently SCD is greater in sub-Saharan African countries such as Cameroon, Republic of Congo, Gabon, Ghana, Nigeria, and Uganda. Uganda has the highest prevalence of sickle cell trait in Africa with a prevalence of about 45% when compared to between 20% and 30% as seen in Cameroon, Republic of Congo, Gabon, Ghana, and Nigeria (WHO, 2012). However, Nigeria has the highest burden of SCD, not only in Africa, but in the world, with about 150,000 children born annually with SCD (Akinyanju, n.d.; Anie, Egunjobi, & Akinyanju, 2010; Vanderbilt University, 2015; WHO, 2006). This high burden of SCD is due to the large population in Nigeria, currently put to over 181 million by the Central Intelligence Agency (2014).

Public Health Impact and Burden of Sickle Cell Disease

Hemolysis (self-breakdown of red blood cells) due to the short life span of the sickled red blood cells results in anemia in patients with SCD is one of the major complications of SCD (CDC, n.d.; Johns Hopkins University, n.d.; NHLBI, 2012). Also, the characteristics of the sickled red blood cells make it easy for the cells to block the flow of blood and oxygen through the body, leading to several complications (CDC, n.d.; Johns Hopkins University, n.d.; NHLBI, 2012). The main complication of SCD due to the obstruction of blood flow is vaso-occlusion and hemolysis (Platt et al., 1994; Rees,

Williams, & Gladwin, 2010). Other common complications of SCD include pain crisis, splenic and renal dysfunction, acute chest syndrome, cerebrovascular complications, priapism, infection, neurologic complications, and acute exacerbations of anemia (Ballas et al., 2010; Platt et al., 1994; Rees et al., 2010). All of these complications lead to early mortality and morbidity in patients with SCD.

People with SCD have a lower average life expectancy when compared with people without this disorder. In the United States, the odds of surviving beyond 70 years, is less than 30% (Platt et al., 1994). In the 1970s, the average life expectancy for people with SCD was about 14 years (NHLBI, 2002; Simon, 2013). However, by 1994, the average life expectancy of patients with the homozygous form of SCD was put at 42 and 48 years for male and female respectively (Platt et al., 1994). This low average life expectancy is significantly influenced by mortality in childhood for SCD patients. The average life expectancy for SCD patients has improved over the years. The average life expectancy of SCD patients is currently put at over 50 years of age. The improvement in survival for SCD patients has been attributed to improvement in the mortality rate in early childhood due to early diagnosis, good follow-up/supportive care, and parental health education (Lanzkron, Carroll, & Haywood, 2013; Quinn, Rogers, McCavit, & Buchanan, 2010; Yanni, Grosse, Yang, & Olney, 2009). In the developing countries, there is inadequate information on the average life expectancy or survival of patients with SCD. However, about 50%-90% of children with SCD have been estimated to die before the age of 5 years in developing countries (Grosse et al., 2011).

Though a global reduction in the under-5 mortality rate due to SCD has been recorded, the mortality rate in children under 5-years-old with SCD continues to be relatively high in Africa (Makani et al., 2011; Ware, 2013). This high under-5 mortality rate due to SCD seen in Africa in relation to the low mortality rate seen in the developed countries has been attributed to inadequate diagnostic facilities in the developing countries, a lack of routine newborn screening for SCD, a lack of an evidence base to support the introduction of some interventions, high mortality in children with SCD due to plasmodium falciparum infection in the developing countries, and suboptimal SCD management and care in developing countries (Ansong, Akoto, Ocloo, & Ohene-Frempong, 2013; Galadanci et al., 2013; McAuley et al., 2010; Obaro 2009; Rahimy et al., 2003). These factors, coupled with the lack of SCD management/control programs in some developing countries, are attributed to the high under-5 mortality rate in SCD seen in African countries.

Apart from the impact of SCD on childhood mortality, SCD also affects the quality of life of patients with this disorder. SCD affects the psychosocial well-being of the child, as well as affect the emotional well-being, school attendance, and school activities of the child (Anie et al., 2010; Barakat, Patterson, Daniel, & Dampier, 2008a; Dampier et al., 2010; Menezes, Len, Hilário, Terreri, & Braga, 2013). SCD also carries its toll on the family. Having a child with SCD carries its psychological and socioeconomic implications for the primary caregiver and the family (Adegoke & Kuteyi, 2012; Barakat, Patterson, Tarazi, & Ely, 2007; Brown et al., 2010; Olagunju, Olaogun, Afolabi, & Adereti, 2014; van den Tweel et al., 2008). The recurrent complications and

rehospitalizations interfere with the caregiver's time, thereby affecting the caregiver's work and finances.

SCD also carries its economic toll on countries with high prevalence of SCD. Due to the complications of this disease, patients with SCD have high rates of health care use and rehospitalizations, with the rehospitalization rates highest for patients with public health insurance (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Kauf, Coates, Huazhi, Mody-Patel, & Hartzema, 2009). The average total cost of care per patient month across all patients with SCD is about 1,946 dollars, with the total cost of care increasing with age. However, the amount of cost attributable to SCD is highest in the 10 to 19 years age bracket, followed by the 0 to 9 years old bracket (Kauf et al., 2009). From infancy to 9 years of age, the total cost of health care due to SCD was reported to be about 892 dollars per patient month (Kauf et al., 2009). In the United States, about 488 million dollars were spent on hospital costs and hospitalizations of patient with SCD in 2004 (Steiner & Miller, 2006). The burden of these hospital costs were carried majorly by public payers of health care, with 66% of this cost being paid by Medicaid and 13% being paid by Medicare (Steiner & Miller, 2006). In 2005, about 335 million dollars was estimated to be spent due to SCD-attributable medical expenditures in children with SCD in the United States (Amendah, Mvundura, Kavanagh, Sprinz, & Grosse, 2010). In Nigeria, the average health care cost of pediatric SCD has also been reported to be high, with parents spending a high percentage of their income on health care for their children with SCD (Abubakar, Lawan, & Bako, 2012; Adegoke, Abioye-Kuteyi, & Orji, 2014). In contrast to developed countries where the cost of health care is borne by public insurance,

a majority of the cost of health care is borne out of pocket, leaving a financial strain on the caregivers of children with SCD.

Management of Sickle Cell Disease

SCD is an inherited disease; hence, preventing this disease is complicated by several ethical controversies. The primary prevention of SCD is premarital screening and genetic counseling which advises a person carrying the sickle cell trait not marry another carrying the sickle cell trait. Premarital screening and genetic counseling is the best way to prevent having a child with SCD, and this approach have been successfully used in reducing the burden of SCD in Middle Eastern countries (Memish & Saeedi, 2011). However, marriage decisions do not change significantly even after genetic counseling (Alswaidi et al., 2012). Also, policies aimed at preventing marriage between persons carrying the sickle cell gene have been termed discriminatory and against human rights (Akinyanju, n.d.). Another strategy employed in preventing SCD is prenatal diagnosis and abortion if positive for SCD. Terminating a pregnancy positive for SCD will help reduce the incidence, and inadvertently, the burden of the disease; however, this strategy is affected by different ethical concerns (Edwin, Edwin, & Etwire, 2011; Fadare, 2009). Though the acceptability of this strategy has been reported among pregnant women with genetic disorders in their families (Ahmed, Ahmed, Sharif, Sheridan, & Taylor, 2012; Gilani et al., 2007; Hewison et al., 2007), prenatal diagnoses and abortions have not been widely accepted among at-risk pregnant women in developing countries like Nigeria (Durosinmi et al., 1995; Durosinmi et al., 1997; Edwin et al., 2011). The health care community in Nigeria does not accept prenatal diagnosis and abortion for preventing

SCD (Adekanle & Adeyemi, 2013; Adeola Animashaun, Nwodo, & Njokanma, 2012; Adeyemi & Adekanle, 2007). With the problems facing the primary prevention of SCD, it is vital to explore secondary strategies in reducing the burden of sickle cell disease.

Secondary prevention of SCD includes management strategies to ameliorate morbidity and mortality due to SCD, as well as to improve the quality of life of persons carrying this disease. The secondary management of SCD includes the use of curative therapy, supportive management, symptomatic management, preventative management, and abortive management (Ballas et al., 2012). The main curative therapy for SCD is the use of stem cell transplantation. Though this method was first successfully used for the treatment of SCD in 1984, the risk benefit consideration of stem cell transplantation, the infrequency of human leukocyte antigen (HLA) identical sibling, the scarcity of matched-unrelated donors, and the high cost of this procedure in the face of lack of government-funded programs for this treatment limit the use of this therapy (Horwitz, 2011; Shenoy, 2013). Other secondary prevention strategies used in the management of SCD includes palliative care, comprehensive clinical care or holistic care, and self-care management.

Palliative care and comprehensive clinical care are similar in that both management strategies involve using an encompassing management system consisting of the SCD patient, different health care practitioners needed for the management of SCD, as well as the families of the patient with SCD (Akinyanju, Otaigbe, & Ibidapo, 2005; McClain & Kain, 2007; Okpala et al., 2002; Rahimy et al., 2003; Wilkie, Johnson, Mack, Labotka, & Molokie, 2010). Both palliative care and comprehensive clinical care focuses on the supportive management, symptomatic management, preventative management,

and the abortive management of SCD. Both management strategies involve the involvement of health care givers in managing and caring for patients with SCD (Akinyanju, et al., 2005; McClain & Kain, 2007; Okpala et al., 2002; Rahimy et al., 2003; Wilkie, et al., 2010). Both palliative and comprehensive care also depends on self-care management by the patients, a majority of the resources used in palliative and comprehensive clinical care requires the patients to adhere to this resource at home.

Self-care management of SCD, unlike palliative and comprehensive clinical care, focuses more on supportive and preventative management of SCD. This management strategy involves the use of resources necessary to prevent SCD complications, as well as developing coping behaviors necessary to improve the psychosocial conditions of people with SCD (Ballas, 2010; Jenerette & Murdaugh, 2008). Though self-care management of SCD is carried out at home without the presence of a health care giver, an initial recommendation or guidance is still required by a health care practitioner (Clark, 1991). Because self-care management does not require the presence of a health care practitioner or require visitation to a health care facility this management strategy is more cost effective for patients with SCD and particular desirable and applicable to developing countries and low-income populations.

Self-care management has been shown to improve the quality of life and to reduce morbidity in patients with SCD (Jenerette, Brewer, & Leak, 2011; Jenerette & Murdaugh, 2008; Tanabe et al., 2010). Nonetheless, patients with SCD may lack adherence to self-care management resources. Although studies have explored the factors that influence, as well as barriers to adherence to some of the resources in SCD self-care management,

these studies have been few, and the scope of these studies has been limited in respect to caregivers of children with SCD. Most of these studies are not recent studies published in the last 5 years. Some of the studies on adherence with SCD self-care management resources were limited to assessing compliance/adherence and the factors influencing compliance to a single self-care management resource (Bitarães, de Oliveira, & Viana, 2008; Elliot, Morgan, Day, Mollerup, & Wang, 2001; Thornburg, 2010; Witherspoon & Drotar, 2006). Other studies only described the factors influencing or barriers to adherence without assessing if there was a significant relationship between the identified factors and adherence (Modi et al., 2009; Patel et al., 2010). A discussion of these studies, along with literature on medication required for use in pediatric SCD cases, effects of these medications on the quality of life of children with SCD and factors influencing, as well as barriers to compliance, with SCD self-care management are presented in Chapter 2.

Problem Statement

Palliative care, comprehensive clinical care, and SCD self-care management have been shown to improve health outcomes in patients with SCD (Jenerette, Brewer, & Leak, 2011; McClain & Kain, 2007; Rahimy et al., 2003). Self-care management involves the use of home-based management to prevent complications of SCD, and it is the only management of SCD that does not require the use of a health care facility or health care practitioner. Even palliative care and comprehensive clinical care requires some form of self-care management by the patient. Compliance to self-care management resources continues to be a problem, especially among parents or caregivers of children

with SCD (Elliot et al., 2001; Witherspoon & Drotar, 2006). In most of these studies, the researchers only looked at one or two aspects of the resources constituting self-care management, but did not look at all the resources of self-care management as a whole. In some other studies, the researchers did not evaluate if there was a significant association between the identified factors that influence compliance or barriers to compliance.

In Nigeria, the psychosocial well-being of the parents significantly impacts the well-being of children with SCD (Adegoke & Kuteyi, 2012; Ohaeri & Shokunbi, 2002; Tunde-Ayinmode, 2011). However, there is little literature on the knowledge of SCD management in the caregivers of children with SCD in Nigeria. There is also a paucity of information on the different factors that promote compliance to SCD self-care management resources among parents/caregivers of children with SCD in Nigeria (Roberts, Izuka, Ekanem, & Mabogunje, 2013).

Purpose of the Study

The purpose of this quantitative study was to examine the knowledge and usage of different SCD self-care management resources among parents/caregivers of children with SCD in Lagos, Nigeria. The aim of this study was also to investigate the level of compliance with the resources of SCD self-care management among the study population and to evaluate the factors that promote compliance with SCD self-care management resources, as well as potential barriers to the usage of SCD self-care management. In this study, such factors that served as independent variables included knowledge, social support, self-efficacy, vulnerability factors, and parental attitudes and parental health

beliefs of caregivers of children with SCD, while the dependent variable was compliance with self-care management resources.

Research Questions and Hypotheses

RQ1-Quantitative-Descriptive: What is the proportion of parents/caregivers of children with SCD in Lagos, Nigeria, that has adequate knowledge about SCD self-care management?

a. What proportion of parents/caregivers of children with SCD has adequate knowledge of SCD, its complications, and ways of preventing complications?
b. What proportion of parents/caregivers of children with SCD has adequate knowledge of SCD self-care management?

RQ2-Quantitative-Inferential: Is there a relationship between the knowledge of self-care management and compliance with the regimen in children with SCD?

$H_0$2: There is no significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

H_a2: There is a significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

RQ3-Quantitative-Inferential: What are the main predictors or barriers to compliance with SCD self-care management among parents/caregivers of children with SCD?

$H_0$3: No statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

H_a3: Statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

Theoretical Framework

The theoretical frameworks for this study were the health belief model (HBM) and the theory of self-care management for sickle cell disease (SCMSCD). According to the HBM, perceived susceptibility, perceived severity of disease, and perceived benefit of a health recommendation may significantly influence the cue to action (Strecher & Rosenstock, 1997). With the underlying concept of personal beliefs, disease perception, and perception about disease prevention influencing the health behavior of a person (Glanz & Bishop, 2010), HBM provides an insight into the influence of parental health beliefs and parental perception of disease severity on compliance with SCD self-care management resources (Elliott et al., 2001). Also according to SCMSCD, vulnerability and health need factors may affect the use of self-care management resources negatively, and self-care management of SCD mediates positively the association between health outcomes and vulnerability factors (Jenerette & Murdaugh, 2008). SCMSCD provided insight into the relationship between vulnerability factors and their influence on the use of self-care management in parents/caregivers of children with SCD.

Nature of the Study

The nature of this study was a quantitative research method, specifically a cross-sectional study. Quantitative research methods can be used to explain a phenomenon by collecting numerical data that can further be analyzed using statistically methods, and this

research method can also be used to determine the relationships among different variables (Creswell, 2009a). Quantitative research was in line with the primary focus of this study and was used to explain, numerically, the knowledge and use of SCD self-care management in my study population (Creswell, 2009a). It was used to identify the variables that best predict compliance to SCD self-care management and the relationship between these variables.

A cross-sectional survey design was used to assess the knowledge of SCD self-care management among caregivers of children with SCD and also to identify the best predictors of compliance, as well as barriers to compliance in this study population. One strength of the cross-sectional study is that data can be collected at a single point in time and it does not necessitate lengthy follow-up processes (Aschengrau & Seage, 2008). Apart from this, cross-sectional surveys can be used to explore the relationship between multiple variables in a single study, which also makes it possible to generalize the characteristics of the study population to a much larger population (Creswell, 2009a; Frankfort-Nachmias & Nachmias, 2008). An interviewer-assisted questionnaire was used to collect information on the dependent variable, adherence, as well as the independent variables of caregiver knowledge, health beliefs, religiosity, self-efficacy, social support, and vulnerability factors. Statistical analysis was used to evaluate the relationship between these independent variables and compliance with SCD self-care management resources.

The signs and symptoms of SCD most times manifests at the age of 6 months and above, and 75% of SCD patients have been said to be diagnosed before the age of 5 years

(Adekile & Adeodu, 2007; Akodu, Diaku-Akinwumi, & Njokanma; 2013; Serjeant, 2013). Mortality due to SCD is highest in the first 5 years of life. In Nigeria, 50% of children with SCD have been said to die before the age of 5 (Obe, 2011). According to the results of previous studies, simple interventions involving the use of prophylaxis penicillin, pneumococcal vaccination, and parental education have been used to significantly reduce mortality due to SCD among children younger than 5 (Quinn, Rogers, McCavit, & Buchanan, 2010; Yanni, Grosse, Yang, & Olney, 2009). Hence, the target population for this study was primary caregivers of children with SCD attending sickle cell clinics and sickle cell support groups in Nigeria and having a child aged 5 years and below with SCD. A convenience sampling technique was used to select sickle cell clinics and sickle cell foundations that were included in the study, as well as to recruit participants into the study.

Definition of Terms

Adherence/Compliance: This is the degree to which a patient follows the use of a regimen according to the recommendations of the health care provider. As an example, adherence is 100% if a patient is required to take a regimen twice a day, and the patients takes the pill twice a day. If the patient is required to take a regimen twice a day, and he or she takes it once a day, this can be said to be 50% adherence (Dunbar-Jacob, 2004).

Parental health belief: This refers to the belief of the parents on the perceived seriousness of SCD complications, perceived susceptibility to complications in their child with SCD, and the perceived benefit of the management regimen in preventing complications in their child with SCD (Elliott et al., 2001).

Parental knowledge: This refers to parents knowing about management regimen that can be used to prevent complications of SCD in children and the recommendations for SCD self-care management.

Religiosity/Spirituality: This refers to the belief that the human experience is under control by a supreme being that increases the ability to rise above the experiences of life and have a greater sense of the purpose of life and satisfaction (Cooper-Effa, Blount, Kaslow, Rothenberg, & Eckman, 2001).

Self-care management: This refers to all actions and coping strategies, carried out at home, and needed to take part in therapeutic behaviors targeted at preventing health complications, improving psychosocial conditions, and maintaining health (Ballas, 2010; Jenerette & Murdaugh, 2008).

Self-efficacy: This refers to the belief that an individual has the ability to perform the daily activities required for a child with SCD. Self-efficacy encompasses all of the skills necessary to manage the complexities of having and successfully managing a child with SCD (Edwards et al., 2000).

Sickle cell disease: This refers to all inherited blood disorders in which the red blood cells contains sickled hemoglobin, whether in the homozygous form (Hemoglobin SS), or as a double heterozygote (Hemoglobin SC and Hemoglobin SB; Behrens & Cymet, 2000; Sergeant, 2013).

Social support: This refers to all available support, whether positive words, financial/other aids, and encouragement that are available to the caregiver of the child with SCD (Jenerette & Murdaugh, 2008).

Vulnerability factors: These refer to predisposing factors that increase the probability of experiencing poor health. These factors may include sociodemographic factors such as age, education, employment and income (Shi, 2001).

Assumptions

The main assumption made in this study was that participants answered truthfully about adhering to self-care management resources. This assumption was necessary for this study because self-reported data, which is a subjective source of information, were used to assess compliance. It was also assumed that participants answered questions on their socioeconomic factors truthfully. Another important assumption in this study was that participants understood the questions as it was asked in English and answered to the best of their knowledge.

Limitations

There is no known available list of children within this age range in Lagos, Nigeria. It was, therefore, not possible to use a probability-based sampling for sample selection. Hence, this study may be subject to selection bias due to the use of a convenience sampling technique. Another source of selection bias in this study was that interviews were conducted in the English language. This may result in the selection of participants who understood the English language, which are mostly people with a high level of education or high social status. Also, because an interviewer-administered questionnaire was used to collect data, this study may be subject to response bias and interviewer bias. Interviewer bias was addressed by training all interviewers on interviewing and the importance of putting participants at ease while interviewing. Other

limitations of this study were due to the inherent nature of cross-sectional studies. Hence, causal inferences that the identified factors are responsible for low adherence cannot be made (Aschengrau & Seage, 2008; Frankfort-Nachmias & Nachmias, 2008).

Scope and Delimitations

The study was limited to primary caregivers of children, 5 years and below, with SCD in Lagos, Nigeria that understand the English language. This age limit was chosen because SCD mortality is highest in the first 5 years of life, and about 50% of children with SCD die before the age of 5 in Nigeria (Obe, 2011). The target population was primary caregivers of children with SCD because children 5 years and below would not understand the magnitude of their illness, and the importance of adhering to self-care management regimens, and would need the help of their primary caregiver in ensuring adherence. The English language limitation put in this study was because Nigeria is a multilingual country with about 250 to 400 languages spoken in the country (Ogunwale, 2013). There is a fusion of tribes and culture in Lagos State as the state serves as the center of excellence and commercial activity in Nigeria. Hence, about 65% of the people living in Lagos are nonindigenes (Akande & Salami, 2010; Ogunwale, 2013). This led to the decision to conduct this study in the English Language, which is the lingua franca in Nigeria.

Only the government-owned sickle cell clinic of the Massey Street Children's Hospital and the laboratory of the privately owned Sickle Cell Foundation were used in this study. The Massey Street Children's Hospital is a major sickle cell referral center that caters specifically for children in Lagos, Nigeria. The Sickle Cell Foundation

laboratory is well equipped for different testing for the complications of SCD and serves as a referral center for laboratory tests for SCD patients in Nigeria. Only factors and barriers to the use of supportive and preventive therapies, but not symptomatic management, abortive management, and curative therapies, were assessed in this study. Abortive management and curative therapies were not included in this study because these are aspects of comprehensive clinical care or palliative care that would require the presence of a health care practitioner. All caregivers of children with SCD in the study centers mentioned earlier were used irrespective of tribe. The use of an interviewer-assisted questionnaire helped reduce the nonresponse or number of missing items in the survey.

Significance of the Study

The fourth millennium development goal (MDG) is to reduce childhood mortality. The targets for this goal are to attain an infant mortality of 30.3 per 1,000 live births and an under-5 mortality rate of 63.7 per 1000 live birth by the year 2015 (United Nations Development Project [UNDP], 2013). In 2012, Nigeria recorded an under-5 mortality rate of 94 deaths per 1,000 live births and an infant mortality rate of 61 per 1,000 live births (UNDP, 2013). With SCD-related mortality constituting 8% of infant/childhood mortality in Nigeria (Sunday Trust, 2013), this project may identify factors that will improve compliance to SCD self-care management. This would help in reducing mortality rates in children with SCD, which would inadvertently help Nigeria in moving closer to achieving the MDG4 targets.

Identifying the factors that promote compliance with the self-care management of SCD may help public health practitioners to explore these factors in designing a possible intervention to promote the use of SCD self-care management among caregivers of children with SCD. This would aid in improving health outcomes in children with SCD in Nigeria. The potential social change implications of this study include revealing if there is adequate knowledge of SCD self-care management among caregivers of children with SCD, which would provide the evidence needed by health practitioners in influencing the introduction of education on SCD self-care management in genetic counseling, especially to couples carrying the sickle cell trait before having a child with SCD and after having a child with SCD. The results of this study may also influence the Lagos State government to include a social welfare package for children with SCD among its health welfare programs. Such welfare package will help provide the basic SCD management resources for families that may not be able to afford such resources.

Summary and Transition

SCD remains a global public health problem leading to early mortality and low health-related quality of life in patients with this disease. Self-care management has been shown to improve the mortality rate and quality of life of patients with SCD. However, compliance with self-care management resources remains a problem. In Nigeria, with its high infant/childhood mortality rate, there is a paucity of data on the knowledge and compliance with SCD self-care management among caregivers of children with SCD. A quantitative research design was used in this study to assess the knowledge of SCD self-care management among caregivers of children with SCD and to explore the various

factors that may serve as promoters and barriers to compliance with SCD self-care management. This study provided evidence that would promote educating parents on SCD self-care management in genetic counseling, as well as influence policies that would provide aids for caregivers of children with SCD.

A detailed description of the study is provided in the following chapters. In Chapter 2, a review of existing literature on SCD management resources for pediatric cases and the effect of these resources on the health-related quality of life of children with SCD are given. An evaluation of literature related to the research questions and objectives of this study are provided in Chapter 2. Chapter 3 provides details of the methodology used in this study. This includes a description of the research design, study site and study population, justification of the sample size, measuring instrument and scales, and validity and reliability of the measuring instrument and statistical analysis used in this study. Chapter 4 comprises of information on the study results and statistical findings, while Chapter 5 discusses the result in relation to the existing body of literature and also provides implication for public health practice and recommendations for future research.

Chapter 2: Literature Review

Introduction

The purpose of this study was to evaluate the knowledge of SCD self-care management among caregivers of children with SCD in Nigeria, as well as identify factors that serve as promoters or barriers to compliance. SCD is a disease of public health importance globally. People with SCD have a low average life expectancy when compared with people with normal red blood cells or people with sickle cell trait (Platt et al., 1994). The low average life expectancy has been attributed to early childhood mortality (Platt et al., 1994). Over the years, there has been an improvement in the average life expectancy of people with SCD. Interventions such as early diagnosis of SCD in childhood through newborn screening, good follow-up, supportive care, and parental health education have helped in improving early childhood mortality in pediatric SCD cases and helping to improve the average life expectancy of patients with SCD (Lanzkron et al., 2013; Quinn et al., 2010; Yanni et al., 2009). Developing countries, especially countries in Africa that have the highest burden of SCD, continue to record high under-5 mortality rate due to SCD (Makani et al., 2011; Ware, 2013). In Africa, factors such as a lack of routine newborn screening, inadequate diagnostic facilities, malaria endemicity, inefficient management of SCD, and a lack of SCD control programs are among the factors implicated in the continuous high under-5 mortality rate due to SCD seen in Africa (Ansong, Akoto, Ocloo, & Ohene-Frempong, 2013; Galadanci et al., 2013; McAuley et al., 2010; Obaro 2009; Rahimy et al., 2003).

Primary prevention of SCD include strategies such as premarital screening and genetic counseling, which advises persons carrying the sickle cell trait against marrying another with the sickle cell trait and prenatal diagnosis and abortion of pregnancy if positive for SCD. The primary prevention strategies of SCD have been construed to be unethical and controversial (Edwin et al., 2011; Fadare, 2009). This is because policies preventing marriage between persons both carrying the sickle cell trait have been said to be discriminatory and impeding individual freedom, while terminating a SCD positive pregnancy have also been controversial and greatly unaccepted in developing countries (Adekanle & Adeyemi, 2013; Adeola Animashaun, Nwodo, & Njokanma, 2012; Durosinmi et al., 1997; Edwin et al., 2011)

In the secondary prevention of SCD, several strategies have been put forward in the management of SCD. Among these strategies includes comprehensive clinical care, palliative care, and SCD self-care management. These secondary strategies have been shown to prevent complications in SCD patients and also to reduce mortality due to SCD (Jenerette et al., 2011; McClain & Kain, 2007; Rahimy et al., 2003, Tanabe et al., 2010). SCD self-care management is an integral component of both comprehensive clinical care and palliative care, as both comprehensive clinical care and palliative care requires some form of self-care management where patients are expected to adhere to some medications and other resources at home for these strategies to work effectively. However, compliance/adherence to the recommended self-care management resources continues to be a problem.

For children with SCD who may not understand the concept of self-care management and the importance of compliance with self-care management resources, parental or caregiver's guidance is required. Complying with the resources of self-care management is a problem among caregivers of children with SCD (Bitarães et al., 2008; Elliot et al., 2001; Thornburg et al., 2010; Witherspoon & Drotar, 2006). However, most of these studies were limited to evaluating compliance with a single SCD self-care management regimen. Researchers have also evaluated the factors that promote or impede compliance. Factors such as health beliefs (Atalla, 2005; Elliott et al., 2001), perceived benefit of a regimen (Thornburg et al., 2010), knowledge of SCD self-care management (Elliott et al., 2001; Rahimy et al., 2003), self-efficacy (Adegbola, 2011; Clay & Telfair, 2007; Edwards et al., 2000; Jenerette & Murdaugh, 2008; Jenerette & Valrie, 2010), social support (Jenerette & Murdaugh, 2008; Tanabe et al., 2010), and spirituality (Adegbola, 2011; Cooper-Effa et al., 2001; Jenerette & Murdaugh, 2008; Tanabe et al., 2010) have all been associated with SCD self-care management. However, most of these studies explained these factors in adult patients with SCD, but not in caregivers of children with SCD, while some used a qualitative approach to factors associated with self-care management.

In developing countries like Nigeria, a majority of children with SCD are from poor families that may not have access to quality care or may not be able to afford quality care (Akinyanju, 2006). The use of self-care management will be useful in improving morbidity and mortality in these children, especially considering that self-care management does not require the presence of health care givers or visiting a health care

facility. There is, however, a paucity of information on the knowledge of SCD self-care management among caregivers of children with SCD, and there is also a dearth of research on the factors that serve as promoters or barriers to compliance to SCD self-care management resources among caregivers of children with SCD. In this chapter, information on the literature search engine, key search terms, and literature search strategies will be provided. Information on the theoretical framework of this study will also be given. In addition, a review of literature on the major complications of SCD in pediatric SCD cases, the medications and other resources required for use in pediatric SCD cases, a review of existing literature on the effects of these resources on the quality of life of children with SCD, and a consideration of literature on the variables that influence compliance will also be provided in this chapter

Literature Search Strategy

Literature used in this literature review was from peered-review journals. In accomplishing this literature review, databases related to health sciences, nursing, and psychology were searched. Such databases included the Cumulative Index to Nursing and Allied Health Literature (CINAHL), MEDLINE with Full Text, ProQuest Nursing and Allied Health Source, PUBMED, Science Direct, ProQuest Dissertation and Theses, PsycINFO, and PsycArticles. The Cochrane Database of Systematic Reviews was used to find review articles on adherence/compliance with SCD management therapies. In addition to these databases, Google Scholar was also used to identified and retrieve some literature. The databases PsycTests and Health Psychosocial Instruments were used to identify measuring instruments that have been used in studying different factors or

variables involved in SCD management. Also, Nigeria has a search engine called Search Nigeria. This search engine only retrieves articles and other publications on a topic as they relate to Nigeria. I used this search engine to further identify studies that have been carried out in Nigeria on SCD and SCD management. The reference lists of all retrieved articles were also used to identify additional literature.

The following keywords were used in the literature search. These words were used alone or in combination with Nigeria, Africa, or developing countries. They included the keywords *self-care management of SCD, management of SCD, management of pediatric SCD cases, adherence in SCD, self-efficacy and medication adherence in SCD, Social support and medication adherence in SCD, spirituality and medication adherence in SCD, knowledge of SCD management,* and *parental knowledge and parental health beliefs and medication adherence in SCD.* As there were few recent articles related to SCD and adherence with SCD management, I did not put any restrictions on the search with regards to publication date or location. Hence, a majority of the articles reviewed had publication dates between 2001 and 2010. All articles reviewed were restricted to those published in the English language.

Theoretical Framework

The intricacies surrounding the adoption of health behaviors involves interactions between factors such as individual experience and conditions, social factors, and environmental factors (Institute of Medicine [US] Committee on Health and Behavior: Research, Practice, and Policy, 2001a). Adopting healthy or preventive health behaviors in people is also dependent on individual beliefs, culture, and values (Institute of

Medicine [US] Committee on Health and Behavior: Research, Practice, and Policy, 2001b). In choosing a theoretical framework for this study, it was important to identify theories that were designed to understand the use of preventive health behaviors in people. Theories included in this study pertain to individual perception and beliefs about a health behavior, as well as theories on the influence of social and environmental factors to a health behavior. Phenomenological theories are used by researchers to identify variables related to health behavior and to explain how these variables interact with each other to influence health behavior (Crosby, DiClemente, & Salazar, 2006). Several behavioral models have been developed and are useful in research involving medication adherence. The HBM, social cognitive theory, and the theory of planned behavior are mostly used to predict adherence, while models such as the transtheoretical model of change have been proposed to change patient behavior (Peterson, Tamiya, & Finley, 2003). The purpose of this study was not to change people's behavior, but rather to identify what influences preventive health behavior and predict adherence. Of the above-mentioned theories, the HBM was originally developed to explore preventive health behaviors, but has found additional use as a health-related behavior model in medication adherence (Janz & Becker, 1984). Hence, the HBM was the main theoretical framework for this study.

Apart from the HBM, another theory that was useful in this study was the SCMSCD (Jenerette & Murdaugh, 2008). This theory is a more recent concept regarding the effect of social and environmental factors on preventive health behavior. According to the SCMSCD, self-care management resources help patients with SCD to have control

over their disease, thereby improving the quality of life in such patients (Jenerette & Murdaugh, 2008). Hence, this theory was important in identifying if the variables identified in the SCMSCD contributed to improving compliance with self-care management regimen and the contribution of these variables to the variance seen in adherence.

The Health Belief Model

The HBM is one of the most widely used theories in explaining health-related behavior. The HBM was developed in the 1950s by Hochbaum, Rosenstock, and Kegels (Hochbaum, 1958; Resource Center for Adolescent Pregnancy Prevention [ReCAPP], 2015). The HBM was initially developed to understand the failure of people to take part in health prevention programs or screening programs that were free (Hochbaum 1958; Rosenstock, 1974). HBM is a value expectancy theory in the context of health behavior, with the assumption that people value good health and would want to avoid illness. Hence, people are predisposed to taking action known to prevent illness and take action recommended for promoting health (Strecher & Rosenstock, 1997). The premise of the HBM is that people will take action on their health if they believe that the illness can be avoided and that taking a recommended action will help prevent the illness (ReCAPP, 2015; Strecher & Rosenstock, 1997). These premises of the HBM are the basis of the constructs of the HBM.

Individuals will take action to prevent, screen for, or manage disease conditions if they believe that they are susceptible to the disease condition, if they believe that the disease condition may have possible complications that are severe, if they believe that

taking a particular action or actions would benefit in reducing their getting the disease or complications of the disease, and if they believe that the benefits of taking the action outweighs the risk/barriers (Strecher & Rosenstock, 1997). Based on these different beliefs, the four main constructs of the HBM are *perceived susceptibility,* which refers to individual perception of the probability of getting a disease condition or illness (Strecher & Rosenstock, 1997)*; perceived severity,* which is the belief that a disease can lead to dire consequences; *perceived benefits,* which refer to the belief that taking a course of action can help prevent the disease or reduce seriousness; and *perceived barrier*s, which is the belief about the factors, which may be cost or other inconveniences, of taking action (Strecher & Rosenstock, 1997). The main hypothesis of the HBM is that perceived susceptibility, perceived severity, and perceived benefit significantly influences the readiness to take action (Strecher & Rosenstock, 1997). In the context of this study, the perceived severity of the SCD and the perceived benefit of SCD self-care management regimen will significantly influence compliance to the SCD self-care management regimens.

Over the years, two other constructs have been proposed and researched in the context of the HBM. These are *cues to action* and the construct of *self-efficacy* (Janz & Becker, 1984, Rosenstock et al., 1988). Cues to action refer to factors that activate or triggers action in people (Strecher & Rosenstock, 1997). The concept of self-efficacy refers to the belief that a person can confidently carry out an action that is required to produce results (Bandura, 1977). The concept of self-efficacy was first introduced by Bandura (1977). This construct was incorporated into the HBM to increase its

explanatory power, especially as related to the use of the HBM in studying behavioral changes associated with the management of chronic diseases (Rosenstock, Strecher, & Becker, 1988). In addition to these constructs, the HBM identifies the influence that other variables such as demographic, social, and psychological factors have on disease perception and on health-related behaviors (Strecher & Rosenstock, 1997). Another hypothesis of the HBM is that sociodemographic factors directly influence perceived susceptibility, perceived severity, perceived benefits, and perceived barriers. Therefore, sociodemographic factors indirectly influence health behaviors (Strecher & Rosenstock, 1997).

HBM originated in the 1950s when Hochbaum (1958) posited the construct of the HBM, perceived belief and perceived susceptibility, in explaining the failure of people to participate in a free tuberculosis screening program. Hochbaum identified that there is an association between perceived susceptibility and perceived benefits and taking a health-related action. Since then, the use of the HBM has been extended to study the health behavior of people in response to symptoms (Janz & Becker, 1984) and compliance of people to medications and other medical regimens (Becker & Rosenstock, 1978; Molfenter, Bhattacharya, & Gustafson; 2012; Park et al., 2010). Park et al. (2010) studied the factors that influence adherence to medications in elderly patients with diabetes mellitus. Park et al. showed in their study that a higher perceived severity and self-efficacy had higher medication adherence with an odds ratio of 2.936 and 4.040 respectively. Those who reported lower perceived barriers also showed a significantly higher medication adherence (Park et al., 2010). Similarly, Mann, Ponieman, Leventhal,

and Halm (2009) identified that disease beliefs (perceived severity) and medication beliefs (perceived benefits and perceived barriers) were significant predictors of medication adherence in patients with diabetes. Health beliefs continue to pay an important role in health behaviors related to medication adherence (Molfenter et al., 2012).

The HBM has also been used to study compliance in SCD. Elliot et al. (2001) used the HBM to assess the influence of parental health beliefs of parents of children with SCD on adherence with prophylaxis penicillin. Elliot et al. showed that perceived barriers were associated with low adherence, and parental health beliefs were responsible for 38% of the variance seen in the adherence to prophylaxis penicillin in pediatric SCD. Witherspoon and Drotar (2006) did not adequately describe HBM constructs in their study. However, Witherspoon and Drotar used a modified Children's HBM and assessed perceived barriers and perceived benefits of the medication using a Beliefs About Medication Scale (BAMS). Caregiver's medication beliefs accounted for about 50% of the variance seen in medication adherence as reported by Witherspoon and Drotar. The relationship between caregiver's medication beliefs and medication adherence was significant with $p < .01$ (Witherspoon & Drotar, 2006). The HBM was used as a theoretical framework in this study to provide an understanding of the influence of parental health beliefs on the different self-care management regimen, parental beliefs about the severity of SCD, parental perception of the benefits of self-care management, as well as the influence of these parental beliefs on adherence with SCD self-care management.

The Theory of Self-Care Management for Sickle Cell Disease (SCMSCD)

SCMSCD is a middle range theory developed by Jenerette (Jenerette & Murdaugh, 2008). This theory was based on a modification of the theory of self-care management for vulnerable populations. The main hypothesis of the SCMSCD is that vulnerability factors such as demographic factors (age, income, education, and employment status), and health need factors negatively affects self-care management and health outcomes. Another hypothesis of this theory is that self-care management resources positively mediate the relationship between health outcomes and vulnerability factors (Jenerette & Murdaugh, 2008). SCMSCD is not applied much in existing literature. However, this theory was tested in the study by Jenerette and Murdaugh (2008). Jenerette and Murdaugh showed that self-care management resources, such as assertiveness, self-efficacy, coping behaviors, social support, self-care ability, self-care actions, and communication skills does not mediate the relationship between vulnerability factors and health outcomes, thereby disproving one of the initial hypotheses of the SCMSCD. This theory was used to augment the HBM in my study, and it provided some additional variables whose mediating effect or interactions with the relationship between the constructs of the HBM and medication adherence can be assessed.

In the following headings, I will review the major complications of SCD in children as seen in the existing body of literature, as well as the main complications in pediatric SCD cases in Nigeria. I will also review the recommended actions for managing these complications. A review of how these actions have been shown to improve the

quality of life in children with SCD will be provided. Additionally, I will review literatures that focused on adherence to pediatric SCD management regimen, as well as review the literature on the key variables associated with self-care management, as well as adherence.

Complications of SCD

The features of the sickle cell gene of being rigid and inflexible are the major cause of complications seen in SCD (NHLBI, 2012; Stuart & Nigel, 2004). Vaso-occlusion and hemolysis are the main pathological processes leading to the clinical manifestations of SCD (Booth, Inusa, & Obaro, 2010). Vaso-occlusion occurs when the sickle cells form heterocellular aggregates with themselves, and with other cells such as leukocytes, and platelets. After forming these aggregates, they adhere to the lining of the blood vessels, thereby blocking the blood vessels. The adherence of the aggregated blood cells to the blood vessels leads to the elicitation of an immune response resulting in pain which is the primary complication of SCD (de Montalembert, 2008; Stuart & Nigel, 2004). The blocked blood vessels can also lead to the reduction of blood flow and, inadvertently, oxygen delivery to organs such as lungs, brain, kidneys, and spleen, thereby leading to secondary complications of SCD (de Montalembert, 2008; Stuart & Nigel, 2004).

Another primary complication in SCD is hemolysis that leads to anemia. This is caused due to the short lifespan of the sickled red blood cells, and its susceptibility to breakdown by autoimmune response (Booth et al., 2010; Ballas et al., 2010). One of the organs affected by vaso-occlusion is the spleen. A series of biological processes in the

spleen renders it hyposplenic or asplenic, which makes it difficult for the body to mount a rapid immune response to fight infection (Booth et al. 2010). Infection is, therefore, another major complication seen in SCD. According to Ballas et al. (2010), the major complications in SCD include hemolytic anemia and its sequelae (such as hyper hemolytic episodes, acute splenic sequestration, and aplastic crises), pain syndromes, and other complications affecting major organs.

Complications of SCD in Children

Researchers have studied the main complications in children with SCD leading to morbidity and mortality. Gills et al. (1995) explored the most common clinical events in the first decade of life of infants with SCD. Painful crises and acute chest syndrome where the most common and frequent complications reported in a cohort of 694 infants with SCD. Infection was the third most common reported complication. Infection was, however, the common cause of deaths in children with SCD in this study (Gills et al., 1995). Similarly, Miller et al. (2000) followed 392 children with SCD from infancy to 10 years in their study. Miller et al. found that three main SCD complications were significant predictors of adverse outcomes in children with SCD. These complications are pain crises (in the form of dactylitis), severe anemia, and leukocytosis (low leukocyte count that may or may not be due to infection). Miller et al. predicted that having dactylitis in the first year of life leads to a relative risk of 2.55 of having an adverse outcome. Having severe anemia can result in an adverse outcome with a relative risk of 2.47, and having leukocytosis even in the absence of infection can lead to an adverse outcome with a relative risk of 1.80 (Miller et al., 2000). Though this model for

predicting adverse outcome in SCD children has been faulted to have low specificity and sensitivity, it still showed that pain crisis, severe anemia, and infection are some of the common and most significant SCD manifestation in children (Quinn et al., 2008).

Other researchers also identified pain crises, infection, and hemolytic anemia as the most common clinical manifestation in children. In Brazil, Filho et al. (2012), with their study on children with SCD in Rio de Janeiro, corroborated that pain crises, infection and hemolytic anemia were the most common clinical manifestation in children with SCD. The study by Filho et al. showed that infection was the most common clinical manifestation, occurring in about 89% of the study participants, and splenic sequestration occurred in about 48% of the children. Filho et al. also showed in their study that acute splenic sequestration was the most common cause of relapse in the children, with 63% of the children experiencing more than one episode. In general, the most prevalent SCD clinical manifestation in this study includes painful episodes (2.9 events/child), infection (2.3 events/child) and hemolytic crises (2.1 events/child; Filho et al., 2012).

Researchers in Nigeria have also studied the most frequent clinical manifestation and complications in children with SCD associated with morbidity and mortality, and like the aforementioned studies, identified that pain crises, hemolytic anemia and infection were significant causes of morbidities. George and Opara (2011) studied hospital data retrospectively for the prevalent clinical manifestations of SCD in patients between 6 months to 18 years in Port Harcourt (located in South-South of Nigeria). According to the results of the study, pain (90%), hyper hemolytic crises (60.4%), and infection where the most frequent clinical manifestations of SCD in children and adolescents (George &

Opara, 2011). The most common infections associated with morbidities in children with SCD in this study are malaria, pneumonia, osteomyelitis, and urinary tract infection (George & Opara, 2011). Similar results were reported in Northern Nigeria by Ambe, Mava, Chama, Farouq, and Machoko (2012). Similar to George and Opara (2011), Ambe et al. (2012) also used retrospective data to assess the clinical manifestation of SCD in children. However, unlike George and Opara, Ambe et al. categorized the clinical features seen based on age. Ambe et al. found that hand- foot swelling (dactylitis) and jaundice were the most frequent symptoms presented between the age of 6 to 11 months, while anemia and vaso-occlusive crises were common at 1 to 5 years. Similar to George and Opara, other causes of morbidity seen in children with SCD in Northern Nigeria included infection in the form of malaria, pneumonia, septicemia, osteomyelitis, and urinary tract infection (Ambe et al., 2012). Brown, Jacob, Lagunju, and Jarett (2013), in their study in South Western, Nigeria, also corroborated the study by George and Opara and Ambe et al. Brown et al. showed in their study that the most frequent clinical presentation of SCD among children with SCD in South Western, Nigeria was vaso-occlusive crises. Brown et al. also identified infection, hyper hemolytic crises, and acute splenic sequestration as other clinical manifestations in children with SCD. The main infections that lead to SCD morbidity in this study were malaria, acute osteomyelitis, pneumonia, urinary tract infection, and septic arthritis. Brown et al. recorded three deaths in their study, one of which was from cerebrovascular accident, one from an adverse reaction to blood transfusion, and one from meningitis. The one death from meningitis

shows that infection also plays a role in SCD mortality, and it would be necessary to identify SCD complications leading to death in SCD patients.

Apart from these conditions that lead to SCD morbidity. Researchers have also studied complications that lead to SCD mortality using autopsy reports. These studies have shown that infection is the major cause of death in SCD cases of all ages. Manci et al. (2003) evaluated 306 autopsy data from SCD patients, accumulated between 1929 and 1996. The study showed that infection was the most common SCD-related cause of death in all ages of SCD accounting for about 33 to 48% of all deaths (Manci et al., 2003). A majority of the infections were located in the respiratory tract, followed by the gastrointestinal tract and genitourinary tract respectively. In children with homozygous SCD (Hb SS), infection-related death is responsible for 80.4% of deaths among children from infancy to two years of age, and 60.6% of deaths from three years to eleven years (Manci et al., 2003). Manci et al. also showed that in children, stroke and sequestration were the next common causes of death in children with SCD.

As also noticed by Manci et al., the mean age of death in SCD increased by the year 1978, this was corresponding to the establishment of several interventions such as newborn screening, prophylactic penicillin therapy, vaccinations, among others, used in improving morbidity and mortality in children with SCD (Manci et al., 2003). However, even in countries where newborn screening programs and other interventions to reduce SCD morbidity and mortality were introduced, the risk of death in children with SCD is still high (Fernandes, Januário, Cangussu, Macedo, & Viana, 2010). Fernandes, et al. (2010) followed a cohort of 1,396 children with SCD from March 1998 to February 2005

in Minas Gerais, Brazil. 78 deaths were recorded during the follow-up period with the main cause of death being infection (38.5%), acute splenic sequestration (16.6%), and other causes (9%). Importantly, the study also showed the presence of unidentifiable deaths in 20.5% of the cases.

In developing countries where most of these prevention programs were not introduced, mortality in children with SCD continues to be on the rise. The causes of mortality in children with SCD in Nigeria also shows similar trends as seen in the study by Manci et al. and Fernandes et al. Autopsy data from Nigeria, as seen in the study by Ogun, Ebili, and Kotila (2014), who evaluated SCD autopsy data between 1991 and 2008, is limited to mortality data in mostly adults with SCD. However, the results followed the trend seen in the study by Manci et al. and Fernandes et al. with infections being the major cause of death, accounting for 78% of deaths in SCD patients (Ogun, Ebili, & Kotila, 2014). Acute chest syndrome and severe anemia were other important causes of death accounting for 37% and 31% of all deaths respectively. The main site of infection was in the respiratory tract accounting for 53.8% of all infections. This was followed by cerebral and urinary tract infection, each accounting for 17.1% of all infections respectively (Ogun et al., 2014).

From the literatures reviewed above, vaso-occlusive pain crisis, infection, and acute exacerbation of anemia are the main causes of morbidity and mortality in children with SCD. Among the precipitating factors for vaso-occlusive crises includes dehydration, infection, fever, stress/fatigue, sudden change in altitude or temperature, exposure to cold and other situations that limit the supply of oxygen to the body, among

other factors (Marlow & Chicella, 2002; de Montalembert, 2008). It is necessary to identify management strategies that would help alleviate these complications in children with SCD.

Self-care Management of SCD in Children

Over the years, several new therapies have been developed due to the understanding of the biology of SCD. These new therapies have helped to improve the quality of life of children with SCD. Though the best management strategy for sickle cell disease and other chronic diseases involves comprehensive clinical or palliative care involving a multidisciplinary group of professionals in disease management (Benjamin, 2008; Claster & Vichinsky, 2003; Lee, Askew, Walker, Stephen, & Artwork, 2012). It is pertinent to note that some form of self-care management is always required even in the case of comprehensive clinical care and palliative care. Considering that there is no cure for SCD, the management of SCD in children involves preventing infection, controlling pain, and managing hydration status (Lee et al., 2012). In the next sections, the SCD management therapies recommended by a healthcare professional, but constitute self-care management will be discussed. Literatures showing the effectiveness of these therapies in children with SCD will also be reviewed.

Prophylactic Medications

The recommended pharmacotherapies used in children with SCD are those that serve as prophylactic medications to prevent infection, anemia, and pain. Examples of prophylactic medications used in SCD management are hydroxyurea, penicillin, folic acid, and in malaria endemic countries, malaria prophylaxis.

Hydroxyurea. This is the only revolution in SCD pharmacotherapy in the past 20 years (Neville & Panepinto, 2011). Hydroxyurea is the only pharmacotherapy approved by the U.S., Food and Drug Administration (FDA) for adult SCD cases, and it is the only available pharmacotherapy able to modify the pathogenesis of SCD (Green & Barral, 2014; Neville & Panepinto, 2011). Hydroxyurea is an antineoplastic agent that is capable of inducing the synthesis of fetal hemoglobin in SCD patients (Lee et al., 2012). Having a high level of fetal hemoglobin in SCD patients reduces the polymerization (aggregation) of sickled red blood cells thereby decreasing disease morbidities in SCD (Green & Barral, 2014). The efficacy and impact of hydroxyurea in adults have been shown in several studies. Charache et al. (1995) carried out a Phase III clinical trial of hydroxyurea using a multicenter study involving 21 sites in the United States and Canada, on the efficacy of hydroxyurea in adults with SCD from 1992 to 1994. The study was a double-blinded, randomized controlled trial of 299 patients, 18 years and above, with SCD, with 152 patients assigned to hydroxyurea treatment and 147 patients to placebo. The hydroxyurea doses given to the treatment group was gradually increased per day depending on body weight and hematological parameters. The result of the study by Charache et al. showed that participants on hydroxyurea had fewer vaso-occlusive crisis and hospitalizations when compared to those in the control group. Hydroxyurea was also shown to reduce the amount of blood transfusions in the treatment group and the number of incidence of SCD morbidities like acute chest syndrome (Charache et al., 1995). Several other studies have also shown the effect of hydroxyurea in adults SCD patients in reducing morbidity and mortality (Steinberg et al., 2003; Voskaridou et al., 2010).

Controversies trail the use of hydroxyurea due to the variability in the response of SCD patients to hydroxyurea, with some patients not responding to it, and also due to the toxicity of hydroxyurea that has been implicated in causing leukemia (Green & Barral, 2014). Studies have shown that the toxicities associated with hydroxyurea are mild and reversible upon stopping the medication (de Montalembert et al., 1999; de Montalembert et al. 2006; Sharef et al., 2013). Several studies have also shown the safety and efficacy of hydroxyurea in children with SCD. Scott, Hillery, Brown, Misiewicz, and Labotka (1996) carried out a pilot study to evaluate the safety and efficacy of hydroxyurea in children. The study by Scott et al. used adolescents aged 10 to 17 years. According to the results of the study, treatment with hydroxyurea improved hematological parameters, reduced hospitalization rates, and showed toxic effects that were acceptable in the study participants (Scott, Hillery, Brown, Misiewicz, & Labotka, 1996). This study helped to show that hydroxyurea could be used in children and adolescents.

The first strong evidence for hydroxyurea in children with SCD was the HUG-KIDS study (Segal et al., 2008). The HUG-KIDS study was a multicenter Phase I-Phase II clinical trial on the safety of hydroxyurea in children. Eighty four children with SCD, aged 5-15 years were included in this study. The study was carried out for 3 years, from 1994 to 1996, and participants were assessed for compliance, toxicity, growth parameters, adverse events, and efficacy of hydroxyurea (ClinicalTrials.gov, 2005). Kinney et al. (1999) using the result of the HUG-KIDS study showed that hydroxyurea increases hemoglobin concentration, mean corpuscular volume, and fetal hemoglobin concentration in the study participants. Kinney et al. also showed that hydroxyurea had

transient toxicities in children, similar to the toxicities seen in adult patients with SCD. Wang et al. (2002) also using the data from the HUG-Kids study reported that there was no adverse effect of hydroxyurea on the growth of children with SCD.

The studies above were based on a Phase I/II clinical trial of hydroxyurea in children. A Phase I/II clinical trial was also carried out on the safety and efficacy of hydroxyurea in infants. This trial called the Hydroxyurea Safety and Organ Toxicity (HUSOFT) trial was a multicenter, prospective, pilot study designed to assess the feasibility of administering hydroxyurea in liquid form to infants with SCD, evaluate the safety and efficacy of hydroxyurea, and to determine the effect of hydroxyurea on organ function (Hankins et al., 2005). The HUSOFT trial included 28 infants with SCD, all less than two years of age, but only 21 infants completed the two years follow-up. Hankins et al. (2005) using data from the HUSOFT study showed that infants on hydroxyurea therapy had a reduced loss of splenic function compare to infants in the control group. Hankins et al. presented the beneficial effects of hydroxyurea on the hematological parameters of infants with SCD, and modest toxicity effects of hydroxyurea were recorded.

The study by Scott et al. and the studies using the HUG-Kids data by Kinney et al. and Wang et al., and the study by Hankins et al. all had the limitations of being open-labeled, single-armed trials (without blinding and without the use of controls). The safety and efficacy of hydroxyurea in children were, nonetheless, shown in these studies. However, due to the inherent limitations of these types of study designs, other studies using a randomized double-blinded clinical trial would be required to improve the

evidence for the use of hydroxyurea in children. Apart from this, Kinney et al. at the end of their study recommended a Phase III trial to evaluate if hydroxyurea would prevent chronic organ damage in children. The BABY HUG trial was subsequently established in order to demonstrate that hydroxyurea prevents organ damage in infants with SCD (ClinicalTrials.gov, 2011; Clinical Trials and Survey Corps, 2008; Thompson et al., 2010). The BABY HUG is a multicenter, double-blinded, randomized, placebo-controlled trial that includes 200 participants, aged nine months to 18 months. Though follow-up of the BABY HUG trial to evaluate long-term toxicity effects of hydroxyurea is still ongoing, the trials have already shown the safety and efficacy of hydroxyurea in infants and young children (Wang et al., 2011). In their article that was based on the result of the BABY HUG hydroxyurea trial, Wang et al. (2011) showed that hydroxyurea significantly reduced the incidence of pain crises, dactylitis, acute chest syndrome, hospitalization rates and transfusion in infants on hydroxyurea therapy compared to the control group. However, there was no significant difference in splenic function (which was the primary objective of the BABY HUG) in both the treatment and control group. Wang et al. concluded that hydroxyurea was safe and efficacious, and should be considered for all pediatric SCD cases. Other studies based on the BABY HUG trial also reported the safety and benefit of hydroxyurea in pediatric SCD cases (Alvarez et al., 2012; Thornburg et al., 2012).

The result of the multicenter study by Charache et al. influenced the approval of hydroxyurea in adult SCD by the FDA in 1998 (Green & Barral, 2014). Though the approval of hydroxyurea has not been extended for pediatric use, this has been attributed

to the unavailability of viable pharmaceutical sponsors (Green & Barral, 2014). The FDA have commissioned a study on the pharmacokinetics of hydroxyurea in a liquid formulation in pediatric SCD (Green & Barral, 2014). This is expected to influence the approval of hydroxyurea in pediatric patients. Nonetheless, it is pertinent to note that despite the lack of hydroxyurea approval in pediatric cases, it is already being utilized for children with severe symptoms of SCD (Neville & Panepinto, 2011). The National Institute of Health (NIH), also identified the fact that further studies were needed in hydroxyurea research, but also identified that currently, the benefits of hydroxyurea outweighed its risk, thereby leading to an NIH approval for the use of hydroxyurea in SCD (Brawley et al., 2008).

Prophylactic penicillin. This has been identified as the most important regimen for the routine management of SCD. Prophylactic penicillin is needed to prevent infection in pediatric SCD cases (NHLBI, 2002). As seen in the studies discussing the causes of morbidity and mortality in children with SCD, infections, particularly infection in the respiratory tract plays a vital role in SCD morbidity and mortality. Infection due to *Streptococcus pneumonia*, *Haemophilus influenza*, and *Salmonella* species have been identified as the major causes of infections in young children with SCD (Booth et al., 2009). One of the earliest evidence for the impact of penicillin prophylaxis in children with SCD was seen in the study conducted by the prophylaxis penicillin study (PROPS) group. This study serves as the foundation for the present guidelines on prophylactic penicillin in children with SCD (Gaston et al., 1986). The study was carried out to

evaluate the effectiveness of prophylactic penicillin in preventing bacterial infections in children with SCD.

The PROPS study was a multi-center, randomized, double-blinded, placebo-controlled trial carried out between 1983 and 1985 on children aged 3 months to 36 months with SCD (Gaston et al., 1986). One hundred and five children were assigned to the penicillin group, and children in this group received penicillin V at a dosage of 125mg twice daily. The placebo group only received 50mg of vitamin C. The study was stopped after 8 months because there was already an 84% reduction in infection in the penicillin group as compared to the placebo group (Gaston et al., 1986). It was concluded that prophylactic penicillin should be administered as a SCD management regimen to children with SCD from the age of 4 months. Another study was carried out by the PROPS group to evaluate the effect of discontinuing penicillin prophylaxis after five years of age. This study, referred to as PROPS II, was also a randomized, double-blinded, placebo-controlled trial (Falletta et al., 1995). The study participants included children who had received penicillin V twice daily for at least two years before the age of five. The intervention group continued to receive penicillin V at 250mg per day while the control group were stopped giving penicillin V by the age of five (Falletta et al., 1995). The results of the study showed four of the participants in the placebo group as against two in the study group suffered from infection, with one participant in each group having penicillin-resistant bacteria strain (Falletta et al., 1995). The authors concluded that penicillin prophylaxis can be discontinued after five years of age. Based on these two PROPS group studies, it was recommended that penicillin prophylaxis (125mg

administered twice daily) be given to pediatric SCD cases from two months of age. This dose should be increased to 250mg, given twice a day, orally, from three years to at least five years of age (NHLBI, 2002; Section on Hematology/Oncology Committee on Genetics). For infants and children allergic to penicillin, erythromycin prophylaxis can be used in its stead (Ndefo, 2008; Section on Hematology/Oncology Committee on Genetics).

Malaria prophylaxis. In Africa, malaria has been identified as one of the causes of morbidity and mortality in children with SCD (Komba et al., 2009; Makani et al., 2010; McAuley et al., 2010). With most countries in West Africa being malaria endemic, and considering the high prevalence of SCD in Africa, it is important to have a form of chemotherapy to reduce the incidence of malaria in children with SCD. According to Fleming (1989), management of SCD in Africa should include malaria prevention. Brousse, Makani, & Rees (2014) posited that SCD patients in Africa should use both insecticide-treated nets and malaria prophylaxis for malaria prevention. Studies have shown that malaria chemoprophylaxis such as proguanil, sulfadoxine-pyrimethamine, and amodiaquine are effective in malaria prevention (Cisse et al., 2006; Eke & Anochie, 2003; Massaga et al., 2003).

Using a randomized, placebo-controlled, double-blinded trial to study the effect of sulfadoxine-pyrimethamine for malaria prevention in children, Cisse et al. (2006) recruited 1136 children, aged two to 59 months. The intervention group was assigned to one dose of artesunate plus one dose of sulfadoxine-pyrimethamine while the control group was assigned to two doses of placebo. The results showed that the use of artesunate

in combination with sulfadoxine-pyrimethamine reduced the incidence of malaria by 6% after three weeks of follow up. Similarly, Massaga et al. (2003) in their clinical trial of the effect of intermittent treatment of amodiaquine on malaria and anemia showed that amodiaquine had a protective efficacy of 64.7% against malaria and 67% against anemia.

In comparing the different malaria chemoprophylaxis drugs, Nakibuuka, Nakiboneka, Ndugwa, and Tumwine (2009) compared the use of intermittent presumptive treatment using sulfadoxine-pyrimethamine and chloroquine for malaria prevention in children with SCD. According to the result of the study, sulfadoxine-pyrimethamine reduced malaria-related hospitalizations by 50% when compared to chloroquine. This difference in malaria hospitalization in the sulfadoxine-pyrimethamine group and the chloroquine group was, however, not significant (Nakibuuka, Nakiboneka, Ndugwa, & Tumwine, 2009). Eke and Anochie (2003) also compared the effectiveness of pyrimethamine and proguanil for malaria prophylaxis in children with SCD. The study was a randomized; placebo controlled; open-labeled study of children, 1 to 16 years, with SCD in Nigeria. In this study, participants were assigned to pyrimethamine (0.5mg/kg weekly) or proguanil (1.5mg/kg daily), or placebo (vitamin C, 7mg/kg daily). The results of the study by Eke and Anochie showed that both proguanil and pyrimethamine reduced parasitemia in children with SCD. However, children on proguanil had lower mean parasite density compared to children in the pyrimethamine group (Eke & Anochie, 2003). There is no general recommendation for a specific malaria prophylaxis in children with SCD, or in children in countries where malaria is endemic. The Federal Ministry of Health in Nigeria recommends the use of indoor residual spraying with insecticides, the

use of insecticide-treated nets, and daily use of the malaria prophylaxis, proguanil, at the dose 25mg daily for children under one year, 50mg for children one to three years, and about 100mg for children three years and above (Nnodu, 2014).

Folic Acid

This is a supplement needed for the production of new red blood cells. The short life span of the sickled red blood cells leads to a depletion of folate stores in SCD patients. Supplementation with folic acid is essential in replenishing the folate stores in the body, and folic acid has been shown to help improve anemia (Stuart & Nigel, 2004). The use of folic acid in SCD patients is controversial. A double-blinded control trial by Rabb et al. (1983) is perhaps one of the first evidence that shows the lack of benefits of folic acid in SCD. Rabb et al. in their study included 117 children with SCD aged six months to four years. The children were randomized to either the folate group or the placebo group (Rabb et al. 1983). Rabb et al. reported that there was no significant difference in the hematological parameters, growth characteristics, infection and other SCD complications in both the folate group and the control group. The researchers also noted mild folate deficiency in the placebo group, and more children came down with dactylitis in the placebo group when compared to the folate group (Rabb et al., 1983). Based on these results, Rabb et al. recommended a review of the policy of the use of folic acid supplementation in children with SCD. In 2008, a double-blinded, randomized, controlled clinical trial was carried out by Hadler, Sigulem, Alves Mde, and Torres (2008) in Brazil. The study included 196 children aged six to 24 months. There was a 14% reduction in anemia prevalence in the folic acid group than in the placebo group.

The researchers, based on the result of their study, reported that there was no significant difference in the incidence of anemia in both the folic acid group and the placebo group in non-anemic children. However, non-anemic children in the folic acid group had a higher level of hemoglobin than non-anemic children in the placebo group. Hadler et al. concluded that folic acid in combination with iron was effective for treating anemia in children.

Folic acid supplementation remains one of the management regimen used in children with SCD, especially in Africa (Fleming, 1989). This is because most countries in Africa are developing countries where there are several cases of malnutrition and under nutrition (Neville & Panepinto, 2011). Folic acid supplementation may, therefore, be necessary to avoid folate deficiency. Folate requirements in patients with hemolytic anemia and SCD are higher than normal due to increased rate of the production of red blood cells. This necessitates the need for supplementary folic acid in this group of people (Incite, 2008). According to Redding-Lallinger as cited in Incite (2008), though folic acid deficiency has not been shown to occur in SCD patients in the U.S., folate deficiency can occur in a short period of time in the case of inadequate food intake or malnutrition. Hence, supplementation with 400 to 1,000 micrograms of folic acid daily would prevent deficiency (Incite, 2008).

Another reason while folic acid supplementation is still crucial in children with SCD in Africa is because of the problem of malaria. Infection with *Plasmodium* species causes a reduction in the hemoglobin levels leading to anemia, and there is also insufficient erythropoiesis during malaria (Chang & Stevenson, 2004; Menendez,

Fleming, & Alonso, 2000). Folic acid can help ameliorate the effects of malaria by replenishing folate stores necessary for erythropoiesis (Ndefo, 2008; Stuart & Nigel, 2004). Folic acid is recommended at a dose of 1mg daily in children with SCD (NHLBI, 2002).

Hydration

One of the factors that may lead to pain crisis in SCD patients is dehydration (de Montalembert, 2008). Episodes of acute pain crisis in SCD patients are first treated with intravenous hydration while oral hydration can be used in mild pain crisis (Okomu & Meremikwu, 2007). Hydration helps to slow down the sickling process while a reduction in the body fluid levels (dehydration) stimulates and uphold the red blood cell sickling process (Okomu & Meremikwu, 2007). According to the Monroe Carell Jr. Children's Hospital at Vanderbilt (n.d.), staying hydrated by drinking plenty of water is one of the best things SCD patients can do to take care of themselves. Fleming (1989) also included being hydrated as one of the principles of the management of SCD in Africa. The Monroe Carell Jr. Children's Hospital at Vanderbilt (n.d.), recommends about two to three 8oz glasses of water for a body weight of 10 pounds, four to six 8oz glasses of water for a body weight of about 25 pounds, and five to eight 8oz glasses of water for a body weight of about 30 pounds.

From the above review, the main principles of management of SCD are the prevention of infection, prevention of infection due to *Plasmodium* species (malaria), prevention of hemolytic anemia and vaso-occlusive crises, prevention of folate deficiency, and prevention of dehydration (Fleming, 1989). The management strategies

of SCD in children, based on the aforementioned principles, and requiring home-based self-care management in children with SCD, include the use hydroxyurea for severe SCD cases, penicillin prophylaxis, malaria prophylaxis in addition to other strategies to prevent malaria, folic acid supplementation, and hydration. In the next sections, the factors that influence compliance with these management strategies will be discussed.

Literature Related to Key Variables and/or Concepts

Adherence/compliance continues to be a problem in the management of SCD. In pediatric SCD cases, parental involvement is required to ensure that children with SCD follow their management regimen as prescribed by the health care professional. Parental involvement has been shown to significantly improve clinical outcomes in children with SCD (Kaslow et al., 2000). Kaslow et al. (2000) suggested that parental education and counseling be carried out to improve the home management of SCD by the parents. Literature showing the problem of adherence to SCD management regimen will be discussed next, as well as the identified factors that influence adherence to SCD management among caregivers of children with SCD.

Adherence/Compliance

Strict adherence to management therapies is necessary for the effective management and prevention of complications in children with SCD. Several studies have reported problems with adherence to medical regimens used for the management of SCD in children. From the first clinical trial of hydroxyurea, Kinney et al. (1999) in the Phase I/II Hug-KIDS study as previously described evaluated compliance with hydroxyurea therapy using pill count. The patients were given enough dosage to cover a two-week

interval, and were expected not to return any pills by the next clinic appointment. Kinney et al. reported that 74% of the patients had full compliance as indicated by not returning any pill at the end of two weeks, 10% of the participants returned less than10% of the hydroxyurea, 10% returned about 25% of the medication, while the remaining 6% returned more than 25% of the medication (Kinney et al., 1999). The HUSOFT trial reported by Hankins et al. (2005) also used return of the medication bottle to assess compliance with hydroxyurea in children with SCD. The researchers recorded about 95% compliance rates with only one participant dropped from the study due to noncompliance. In the BABY HUG trial, adherence to hydroxyurea was also measured based on measuring the content of returned medication bottles (Thornburg et al., 2010b). The measure of adherence was calculated using the formula

$$\frac{\text{Amount consumed}}{\text{Prescribed amount}} = \frac{\text{(amount dispensed)} - \text{(amount returned)}}{\text{(daily dose prescribed)} \times \text{(\# days on dose)}} \times 100\%$$

Based on this, Thornburg et al. reported an 88.9% adherence in the patients (Thornburg et al. 2010b). In another study on adherence to hydroxyurea, Thornburg et al. (2010a) used a cross-sectional study of children with SCD to compare different methods of measuring adherence. The procedures for measuring adherence compared in the study includes number of pharmacy refills, a Modified Morisky Scale, caregiver report, and a visual analog scale (Thornburg et al., 2010a). The researchers reported that adherence to hydroxyurea in children with SCD ranged from 49% to 85% depending on the adherence measure. According to the findings of the study, the estimate of adherence may vary

depending on the adherence measure used and that pharmacy refills provided the lowest estimate of adherence (Thornburg et al., 2010a).

Adherence problems were reported concerning penicillin prophylaxis. In fact, non-adherence to penicillin prophylaxis can lead to dire consequences. Buchanan and Smith (1986) revealed the importance of strict adherence to prophylactic penicillin. The researchers followed 88 children with SCD over a period of 7 years. The authors recorded pneumococcal septicemia in six participants, five of which were not taking their prophylactic penicillin when they got infected. The sixth participant that came up with an infection did so after missing just a few dose of medication (Buchanan & Smith, 1986). Generally, low adherence to prophylactic penicillin has been reported. Teach, Lillis, and Grossi (1998) reported poor compliance with prophylactic penicillin in children with SCD. A 43.1% compliance with penicillin in children with SCD was reported by Teach et al. It was also showed that children younger than five years were more compliant than older children, and children with private health insurance were more compliant than children with public insurance (Teach, Lillis, & Grossi, 1998).

Other researchers have also examined compliance with prophylaxis penicillin in SCD. Berkovitch et al. (1998) examined compliance to prophylactic penicillin prophylaxis in children, 9 to 84 months, with SCD. The researchers used a Medication Event Monitoring System (MEMS) to assess compliance. MEMS is a medication bottle "that monitors the timing and frequency of bottle openings" (Berkovitch et al., 1998, pg. 605). The authors found a variable compliance rate of 1.3 to 98.2% at baseline. The researchers noted that the MEMS device is not without its problems, as the device cannot

be used with medication in liquid formulation, and children may frequently drop the device. The researchers, thus, concluded that measuring medication compliance may be problematic. Elliott et al. (2001) compared pharmacy refills and caregivers report in measuring compliance to prophylactic penicillin in children with SCD. A compliance rate of 12% was reported using pharmacy refills in contrast to 60% compliance as reported by the caregivers. Elliot et al. substantiated the findings of Teach et al. that parents of younger children were more compliant than parents of older children, though this difference was not statistically significant. Also assessing compliance to prophylactic penicillin, Bitarães, et al. (2008) and Witherspoon & Drotar (2006) recorded a 48.1 and 56.7% adherence to prophylactic penicillin, respectively, among caregivers of children with SCD. Studies on compliance to prophylactic penicillin have shown inconsistent compliance rates using different methods. Bitarães et al. (2008) recommends using several methods and not relying on one method of evaluating compliance.

Unlike the other studies discussed above, Patel et al. (2008) assessed adherence to multiple SCD management therapies in their study. The SCD therapies included in their study includes penicillin, hydroxyurea, and folic acid. Patel et al. measured adherence "as the ratio of the number of expected days between refills periods (numerator) and the observed days between refill periods for the patient (denominator)" (Patel et al, 2008, pg. 555). The authors showed adherence rates of 54.9%, 60.5% and 61.3% for penicillin, hydroxyurea, and folic acid respectively.

Parental Health Belief

Parental health and medication belief have been shown to influence compliance with SCD management regimen. Elliot et al. (2001) evaluated the effect of parental health beliefs on medication adherence in children with SCD. The study included 50 mothers of children with SCD with an age range of 6 months to 60 months. Elliott et al. assessed compliance rates with prophylactic penicillin, as well as factors that influenced compliance with prophylactic penicillin. The researchers showed that the perceived burden of obtaining medication refill and remembering to administer the medication were the only significant factors of parental health beliefs that influence compliance, and that parental health beliefs contributed to 30% of the variance seen in compliance (Elliot et al., 2001). Witherspoon and Drotar (2006) reported that parental health beliefs (this time in the form of medication belief) accounted for 50% of the variance seen in adherence to prophylactic penicillin. Similar to Witherspoon and Drotar, Thornburg et al. (2010a) showed that parental beliefs about medication (in this case hydroxyurea) influenced adherence to this medication. The study by Thornburg et al. (2010a) did not use a validated measure to assess parental health beliefs, but the researchers showed in their study that parental wariness of hydroxyurea as a prophylactic measure influenced adherence to this therapy in six children.

Parental Knowledge

Lack of adequate knowledge about SCD management and the importance of complying with SCD management regimen may influence medication adherence. Insufficient knowledge about SCD management in caregivers of children with SCD has

been shown to influence self-efficacy and coping behaviors in children with SCD (Barakat, Simon, Schwartz, & Radcliffe, 2008b). There are contrasting reports of the influence of parental/caregivers knowledge on adherence. Berkovitch et al. (1998) in their study on compliance with prophylactic penicillin found a high compliance rate at baseline. This high compliance rate found at baseline was attributed to health care practitioners reiterating the importance of prophylactic penicillin at the study site, at each clinical visit, which may have increased the knowledge of the parents about the medication and its importance (Berkovitch et al., 1998). However, Berkovitch et al. noted no significant difference between parental knowledge and compliance (Berkovitch et al., 1998). Elliot et al. though did not measure parental knowledge and its relationship with compliance, the researchers, however, noted that parents whose children were often ill were more knowledgeable about SCD management and were generally more compliant (Elliott et al., 2001). Barakat, Smith-Whitley, and Ohene-Frempong (2002) also showed a significant association between SCD knowledge in caregivers of children with SCD, and medical staff rating of medication adherence. Witherspoon and Drotar did not provide the measures used for caregiver knowledge, they, however, noted that caregiver's reported knowledge of infection were strong predictors of adherence. Witherspoon and Drotar also noted that caregivers who had better knowledge of infection reported fewer barriers to adherence to prophylactic penicillin.

Jensen et al. (2005) studied the effect of caregiver knowledge on adherence in children with SCD. The study was a cross-sectional study of parents of children with SCD attending a pediatric sickle cell clinic. A Self-Care Inventory-Sickle Cell (SCI-SC)

scale was used to assess compliance with specific behaviors necessary in children with SCD. Jensen et al. used the Transition Knowledge Questionnaire and the Measure of Knowledge about Sickle Cell Disease to assess parental knowledge of SCD. The researchers showed that there was no linear correlation between parental knowledge of SCD and adherence. The researchers, however, noted that their result only showed that parental knowledge about SCD was not the most significant variable predicting adherence but did not establish the lack of a relationship between parental knowledge of SCD and adherence (Jensen et al., 2005). Using post hoc analysis, Jensen et al. showed that in the case of parents with younger children, those with a high level of general knowledge had better-reported adherence (Jensen et al. 2005). This is in line with the result of the study by Teach et al. and Elliot et al. that compliance was better in younger children than in older children. Oyeku et al. (2013) also studied the influence of parental factors on the use of hydroxyurea in children with SCD. Unlike other studies, Oyeku et al. showed that parental knowledge was associated with hydroxyurea use.

Religiosity/Spirituality

With the complications of SCD and the various management regimens required for self-care, patients with SCD need to develop different methods to cope with their disease. Among the approaches used for coping with SCD includes psychological counseling, hypnosis, and prayer (Cooper-Effa et al. 2001; Cotton et al., 2009). Spirituality has been identified as a coping mechanism, and is recognized as one of the factors that contribute to the management of chronic illnesses (Adegbola, 2011; Cooper-Effa et al., 2001). According to Cooper-Effa, Blount, Kaslow, Rothenberg, and Eckman

(2001), spirituality refers to the belief that human experience is under control by a supreme being that increases the ability to rise above the experiences of life, and have a greater sense of the purpose of life and satisfaction. There are limited studies on the impact of religiosity/spirituality in SCD patients, and most of the available studies focused on the role of spirituality in coping, and to pain measures in adults or adolescents with SCD (Adegbola, 2011; Cooper-Effa et al., 2001, Cotton, 2009). However, none of the available studies considered the role or the impact of religiosity/spirituality to adherence to medication in patients with SCD.

Factors associated with coping and adjustment can be associated with treatment adherence. This is because treatment adherence can be classified as an aspect of coping or adjustment to chronic disease, as suggested by Christiaanse, Lavigne, and Lerner (1989). One recent study supported this view by identifying the impact of religiosity/spirituality on medication adherence in children with cystic fibrosis. Children with cystic fibrosis, similar to children with SCD, require daily health management which involves clearing the airway twice daily, inhaling nebulized medications, oral antibiotics, and nutritional supplements (Grossoehme et al., 2013). In the study by Grossoehme et al. (2013), parents of children, aged three months to 13 years, with cystic fibrosis were interviewed. Grossoehme et al. used grounded theory methodology to evaluate adherence and spirituality in parents of children with cystic fibrosis in this study. The researchers identified that some parents believed there was no relationship between their faith and treatment adherence (Grossoehme et al., 2013). Among parents that had high adherence, Grossoehme et al. showed that high adherence parents believed that they were

empowered by God to take care of their child and they used prayer to change themselves to be adherent, while parents with low adherence posited that they trusted God to take care of their child, and they used prayer to change God (Grossoehme et al., 2013). The use of a qualitative approach in the study by Grossoehme et al. helped to identify parental perception about spirituality and adherence (Grossoehme et al., 2013). However, the limitation of this study is that it cannot be used to infer if there is a significant relationship between spirituality and medication adherence.

Self-efficacy

Self-efficacy is another variable that may influence coping in people with chronic disease. Self-efficacy is the belief that one can successfully carry out an action required to achieve a desired outcome (Clark & Dodge, 1999; Edwards et al., 2000). According to Hawkins (1992), self-efficacy is a predictor of behavior, but not a cause of behavior itself. Hence, self-efficacy is specific for a given behavior (Clark & Dodge, 1999). In this regard, someone with self-efficacy would be confident about successfully taking a medication as prescribed. Clark and Dodge (1999) showed in their study that self-efficacy was a predictor of certain disease management behaviors.

The concept of self-efficacy in SCD patients was evaluated, but none of the studies evaluated whether self-efficacy was a predictor of medication adherence in SCD. Clay and Telfair (2007) studied if self-efficacy was a predictor of adjustment in adolescents with SCD. The researchers reported that self-efficacy accounted for about 50% of the variance seen in adjustment (Clay and Telfair, 2007). Based on the premise that medication adherence is a feature of coping/adjustment, self-efficacy may also

influence adherence. Jenerette and Murdaugh (2008) identified in their study that self-efficacy in combination with factors such as self-care ability, assertiveness, and social support assist in the management and coping of adults with SCD. Harper et al. (2013) studied the effect of parental caregiving self-efficacy on the ability to provide care for their children with cancer. Caregiving self-efficacy in parents influenced their reaction to their children's treatment procedures (Harper et al., 2013).

Social Support

Social support has been shown to play an important role in health behavior, and in adherence to health management regimen. There are limited studies on the role of social support on compliance in SCD patients. Belgrave and Lewis (1994) showed in their study that social support was significantly associated with adherence to health management activities. Belgrave and Lewis also noted that providing social support to patients with SCD and diabetics helps to improve compliance with health activities. Chen, Cole, and Kato (2004) in their review of psychosocial interventions in pain and adherence in SCD showed that social support interventions were important in enhancing SCD care and daily management. Social support has also been shown to improve adjustment and coping behaviors in SCD patients (Chen, Cole, & Kato, 2004; Gold, Treadwell, Weissman, & Vichinsky, 2008). Jenerette and Murdaugh (2008) identified social support as one of the factors that assist adults with SCD manage their disease. However, whether social support influenced medication adherence, which then influenced health outcome, is a phenomenon that is yet to be studied. The contribution of social support to the variance seen in adherence also needs to be studied.

Vulnerability Factors

Factors such as age, income, education, and employment, and even marital status have been identified as sociodemographic variables that can also be referred to as vulnerability factors (Jenerette & Murdaugh, 2008). In the SCMSCD, these sociodemographic factors influence health outcomes in patients with SCD (Jenerette & Murdaugh, 2008). There are also differing results on the influence of sociodemographic factors on medication adherence among caregivers of SCD patients. Elliot et al. showed in their study that demographic variables accounted for 38% of the variance seen in compliance. Among the demographic variables measured were number of adults in the house, availability of a car in the family, and maternal education (Elliot et al., 2001). A significant association was found between number of adults in the house and having a family car, with families having more than one adult in the house, and families having a car having better adherence to prophylactic penicillin, but there was no significant association between maternal education and compliance. Witherspoon and Drotar (2006) measured the same demographic variables as Elliott et al. but did not find any significant relationship between any of these demographic variables and adherence. Likewise, Bitarães et al. (2008) did not find any significant association between medication adherence and number of family members or adults in the family, caregiver's education, age. and gender.

Caring for a child with SCD carries a financial burden on the family. Having a child with SCD carries its psychological and socioeconomic implication on the primary caregiver, and on the family (Adegoke & Kuteyi, 2012; Brown et al., 2010; van den

Tweel et al., 2008). This financial burden may lead to a longer period between refills of medication and may inadvertently affect adherence to medication. Barakat et al. (2002) showed in their study that family income was significantly associated with medical staff rating of adherence (p = .023), and family income accounted for 4% of the variance seen in medical staff rating of adherence. Likewise, Witherspoon and Drotar found in their study that families with employment had better adherence rates than families without employment. Unlike Barakat et al. (2002) and Witherspoon and Drotar (2006), Bitarães et al. did not find a significant association between family per capital income and adherence.

Barriers to Adherence

Several factors have been attributed to the problem of nonadherence to prophylactic medication among children and adolescents with SCD, as well as their caregivers. Among the factors that have been identified as barriers to compliance include forgetting to administer medication, burden of picking up refills due to lack of transportation, among others. Studies' indicate that neglect of medication administration and perceived burden of obtaining medication refills are the primary barriers to adherence seen among caregivers of children and adolescents with SCD (Elliot et al., 2001; Modi et al., 2009; Thornburg, et al. 2010a; Witherspoon & Drotar; 2006). Elliott et al. showed in their study that the perceived burden of picking up refills and forgetting to administer medication were the most significant factors influencing adherence to prophylactic penicillin (p <.01). Elliott et al. reported that families without a household car had less compliance than families with a household car (Elliot et al., 2001). The absence of a

convenient means of transportation affects the perceived burden of picking of refills of the caregivers, and inadvertently impact compliance.

In contrast to Elliott et al. (2001) whose study was limited to adherence to prophylactic penicillin, Modi et al. (2009) covered a wider range of SCD prophylactic medications such as hydroxyurea, medication for pain therapy, vitamins and mineral supplements, oral antibiotics, hydration, and medication for chelation therapy in their study. Modi et al. showed form their findings that forgetting to administer medications was the most reported barrier to adherence to the entire prophylaxis regimen with the exception of chelation therapy. Similar to Elliott et al., the study by Witherspoon and Drotar (2006) also assessed adherence to only prophylaxis penicillin in children with SCD. Witherspoon and Drotar showed that 26.7% of the caregivers in their study reported being busy as a barrier to adherence, 23% reported forgetting to administer medication, 20% reported child falling asleep, and 16.7% reported ran out of medication as barriers to compliance. Overall, forgetting to administer medication and hospital refill of medications has been identified as barriers to compliance with SCD therapy.

Summary and Transition

Vaso-occlusive pain crisis, infection, and anemia are the major complications causing morbidity and mortality in children with SCD (Ambe et al., 2012; Brown et al., 2013; Filho et al., 2012; George & Opara, 2011; Quinn et al., 2008). Over the years, several management recommendations have been provided to help prevent these complications in pediatric SCD. Among the health recommendations include the use of hydroxyurea for severe cases of SCD to prevent vaso-occlusive and pain crisis;

prophylactic penicillin to prevent infections; malaria prophylaxis such as proguanil, amodiaquine/chloroquine, and sulphadoxine-pyrimethamine to prevent malaria in children living in malaria endemic countries; folic acid to prevent anemia;and hydration (Fleming, 1989; Nnodu, 2014). Though the effectiveness of these management recommendations have been shown, adherence to these regimens remains a critical problem, especially in pediatric SCD cases where caregiver's guidance is required (Barakat et al., 2002; Bitarães et al., 2008; Elliot et al., 2001; Thornburg et al., 2010a, Witherspoon and Drotar, 2006).

Adherence to SCD management regimen varies depending on the adherence measures used. Adherence measures such as the Morisky scale, pharmacy refills, caregiver self-report, MEMs, and even medical staff estimates have been shown to provide differing results in the measurement of adherence (Bitarães et al., 2008; Elliot et al., 2001; Thornburg et al., 2010a). Hence, measuring adherence may be problematic. Among the factors that have been shown to influence adherence to SCD management regimen among caregivers of children with SCD includes the health beliefs of the caregivers, caregiver's knowledge of the importance of the management regimen, religiosity/spirituality, self-efficacy, social support, and other sociodemographic factors that serves as vulnerability factors.

Perceived medication beliefs and perceived burdens were the most significant factors of parental health beliefs associated with adherence with penicillin and prophylactic penicillin (Elliot et al., 2001; Thornburg et al., 2010a; Witherspoon and Drotar, 2006). However, a gap still remains which is to identify if these same constructs

of parental health beliefs will be associated with adherence to malaria prophylaxis, folic acid, and hydration, and if these same constructs of parental health beliefs would influence adherence among caregivers of pediatric SCD in Nigeria. There are contrasting results on the influence of parental knowledge of SCD management regimen on adherence with some studies finding a significant association between parental knowledge of SCD management and adherence (Barakat et al., 2002; Elliot et al., 2001; Oyeku et al., 2013; Witherspoon and Drotar, 2006) while others showed no significant association between parental knowledge and adherence (Berkovitch et al., 1998; Jensen et al., 2005). It is pertinent to note that apart from Jensen et al. (2005), other studies did not use a validated method to measure parental knowledge. This is one of the limitations to be addressed in the present study.

Both spirituality and self-efficacy improved coping and adjustment to SCD (Adegbola, 2011; Cooper-Effa et al., 2001, Cotton, 2009; Clay and Telfair, 2007; Jenerette & Murdaugh, 2008). However, there is a gap in the literature on whether spirituality and self-efficacy influences compliance in caregiver's of children with SCD, and if the relationship between these variables and adherence is significant. There is also no information on the influence of social support on medication adherence in SCD, though social support have been associated with adjustment and coping behaviors in SCD patients (Chen et al., 2004; Gold et al., 2008; Jenerette and Murdaugh, 2008). The influence of social support on adherence is a phenomenon that still needs to be studied.

There are also inconsistent results on the influence of sociodemographic factors and adherence. Though sociodemographic factors have been reported to have a

significant influence on adherence (Elliot et al., 2001; Barakat et al., 2002), a majority of the researchers showed a lack of significant association between sociodemographic variables and adherence (Bitarães et al., 2008; Witherspoon & Drotar, 2006). In general, the main barriers to adherence to SCD management reported in the literature are forgetting to administer medication and burden of picking up refills due to lack of transportation, among others (Elliot et al., 2001; Modi et al., 2009; Thornburg, et al. 2010a; Witherspoon & Drotar; 2006).

Given the inconsistencies on the influence of parental knowledge of SCD management on adherence, and the influence of sociodemographic factors on adherence, the question still remains, does parental knowledge of SCD management regimen influence adherence to such regimen, and does sociodemographic factors influence adherence to SCD management regimen? Also, no study has used inferential methods to look at the influence of religiosity/spirituality on medication adherence, self-efficacy on medication adherence, as well as social support on medication adherence among caregivers of children with SCD. I, by this study, aimed to answer the above questions, and also study the relationship between spirituality, self-efficacy, social support, and adherence. The research design and approach used for this study are explained in the next chapter (Chapter 3). A justification of the choice of the study design, determination of sample size, and selection criteria are similarly provided. Finally, I also provide information on the measurement instruments and data analysis.

Chapter 3: Research Method

Introduction

The purpose of this study was to examine if there is an adequate knowledge of self-care management among caregivers of children with SCD in Lagos, Nigeria, as well as to identify the factors that influence (promoters and barriers) compliance with self-care management. These gaps need to be addressed in other to design interventions that will focus on improving adherence to self-care management among pediatric SCD cases, as well as to inform policies that will help remove barriers and improve adherence to self-care management in this population. As the problem being addressed in this study requires measuring adherence rate and making inferences about the relationship between several variables and adherence, a quantitative approach to this study was reasonable.

In this chapter, I discussed and rationalize the research methods used in this study. The discussion in this chapter includes a justification for the research design, study population, sample size, and sampling procedure. Information on the data collection process, measurement instruments, and operationalization of the variables are also provided. I also provide information on the potential threats to validity and the ethical concerns to be considered in this study.

Research Design and Justifications

The purpose of this study was to examine the knowledge of SCD self-care management among caregivers of children with SCD, evaluate adherence to the SCD self-care management regimen, as well as to identify the factors that influence compliance. Hence, the dependent variable in this study was compliance to self-care

management resources. Among the factors that influence adherence that were considered as independent variables in this study were caregiver's knowledge of SCD management, parental health beliefs, religiosity/spirituality, self-efficacy, social support, and sociodemographic factors that may serve as vulnerability factors.

As this study involves identifying factors that influence an outcome (in the case of this study, adherence) and the best predictors of compliance, it was appropriate to use a quantitative approach (Creswell, 2009b). According to Creswell (2009b), a quantitative approach is the best approach to use if a research problem involves identifying variables that influence an outcome and also recognize the best predictors of an outcome. A qualitative approach was not desirable for this study as the problem of adherence was not new, and there are available researches on the factors that influence adherence. The variables to examine in relation to adherence were already known and did not require an exploratory approach to identifying these variables (Creswell, 2009b). As a quantitative approach was adequate to answer the research questions, there was no need to use a mixed-methods methodology for this study, especially in the face of time and resource constraints.

This study was carried out using a cross-sectional survey design. Cross-sectional surveys are exploratory in nature and are usually applied in researches that involve identifying associations or relationships between variables and research that involves generating a hypothesis that can then be tested with an experimental research design (Carlson & Morrison, 2009; Frankfort-Nachmias & Nachmias, 2008). A cross-sectional survey was appropriate for this study because information could be collected from the

study participants at a single point in time (Creswell, 2009a, 2009b). Based on this characteristic of the cross-sectional study design, such study design requires a relative shorter time to carry out, as well as require fewer resources to conduct (Carlson & Morrison, 2009; Frankfort-Nachmias & Nachmias, 2009; Hall, 2008). By using a cross-sectional study, a hypothesis can be generated from the multiple variables assessed, which can advance knowledge as related to the topic through further testing using an experimental study (Carlson & Morrison, 2009).

A cross-sectional study was appropriate not only to answer the research questions, but was suitable for a dissertation study in order to avoid the lengthy follow-up process associated with longitudinal and experimental studies (Aschengrau & Seage, 2008). By using a cross-sectional research design, I was able to avoid problems with loss to follow-up associated with longitudinal and experimental studies (Aschengrau & Seage, 2008). In addition, using a cross-sectional research design makes it easy to collect information on multiple variables in a single study, and it allows for flexibility in the different data collection methods that can be used to collect information in such study designs (Creswell, 2009a; Frankfort-Nachmias & Nachmias, 2008; Hall, 2008). The cross-sectional survey design is not without its limitations, among which includes the fact that using a cross-sectional research design does not allow for the establishment of causal effect between the independent and dependent variables and may also have some limitations in having a low internal validity (Frankfort-Nachmias & Nachmias, 2008; Szklo & Nieto, 2014). A cross-sectional survey design was used to obtain information on

the caregiver's knowledge, caregiver's health beliefs, caregiver's self-efficacy, and adherence to self-care management resources at a single point in time.

Methodology

Information on the target population, inclusion and exclusion criteria for the participants, sample size considerations, sampling and recruitment strategy, and data collection procedures are discussed in this section.

Target Population

The target population for this study was the primary caregivers of children; 5 years and below; with SCD; attending sickle cell clinics in public hospitals in Lagos, Nigeria; or attending sickle cell support groups in Lagos, Nigeria. This population represents a finite population of people that is characterized by a countable sampling unit (Frankfort-Nachmias & Nachmias, 2008). However, there is no available data on the overall population size of this target population in Lagos, Nigeria. Because caregiver support is required by children due to their young age, this fact influenced the choice of the target population in this study.

The inclusion criteria for the sampling was that the caregivers must be the primary caregiver of the child with SCD, must have a child 5-years-old or below the age of 5 with SCD, must be attending a sickle cell clinic in one of the government-owned hospitals in Lagos state or a sickle cell support group, must understand the English language, and must be willing to participate in the study. The age limitation placed on the child with SCD is because SCD manifests its symptoms by the age of 6 months, and about 75% of people with SCD are diagnosed before the age of 5 (Adekile & Adeodu,

2007; Akodu et al.; 2013; Serjeant, 2013). In Nigeria, the percentage of children with SCD that die in the first 5 years of life is still high, estimated at 50% (Obe, 2011). This information influenced the age criteria placed in this study. English language was chosen as the language for this study, despite the fact that there are other local languages in Nigeria, because the English language is the official language in Nigeria. Nigeria is a multilingual country with about 250 to 400 languages spoken in the country (Ogunwale, 2013). Lagos state, where this study was carried out, is a melting point of different cultures and languages in Nigeria, and Lagos serves as the major point of commercial activities attracting people from all over the federation. Hence, about 65% of the people living in Lagos are nonindigenes (Akande & Salami, 2010; Ogunwale, 2013). Based on the problem of multiple languages spoken by people in Lagos state, this study was conducted in the English language which is the lingua franca in Nigeria.

Sampling

With the lack of information on the number of people constituting the above-mentioned target population used in this study, and with the consideration that this target population was small, a probability sampling unit was not adequate to achieve an appropriate sample size (Frankfort-Nachmias & Nachmias, 2008). Hence, a convenience sampling method was used for sampling. One of the inclusion criteria was that the caregivers must be attending a government-owned hospital in Lagos Nigeria. A convenience sampling method was used in selecting the study sites used in this study. The study participants were volunteers attending the Massey Street Children's Hospital

and the Sickle Cell Foundation for their scheduled checkup. A minimum of 100 participants was included in the study.

Sample Size Considerations

In calculating sample size for regression models, several rules have been proposed in estimating an adequate sample size (Van Voorhis & Morgan, 2007). Among these rules are the ones recommended by Green (1991) for the smallest acceptable sample size for testing the overall fit of a regression model and in testing the contribution of individual predictors to the model. Green recommended using a sample size greater than $50 + 8m$ (where m is the number of predictors) in testing the overall fit of the model and $104 + m$ in testing the contribution of each predictor to the model. Considering that regression models require testing both overall fit and contribution of individual predictors, Green suggested using the formula that will give the largest sample size, which is $104 + m$. Field and Miles (2010) posited that these rules oversimplify the problem of calculating the required sample size for regression analysis.

In order to calculate an adequate sample size, the statistical power, significant level (alpha), and effect size were considered (Sheperis, n.d.; Sullivan, 2012). The statistical power is the probability of correctly rejecting a false null hypothesis, alpha is the probability of failing to reject a true null hypothesis, and the effect size is a measure of the magnitude of the expected relationship between the variables (Sheperis, n.d.; Sullivan, 2012). The acceptable benchmark of a statistical power of 80% (0.8), and a significant level of 0.05 was used (Cohen, 1988; Sullivan, 2012). For regression models, the appropriate effect size measure is Cohen's f^2 (Sheperis, n.d.). f^2 is a function of R^2,

which is the square of multiple correlations. f^2 is given as R^2 divided by 1 minus R^2. There are differing reports on the amount of variance (R^2) contributed by various factors to adherence in caregivers of children with SCD. Elliott et al. (2001) reported that 53% of the variance seen in adherence to prophylactic penicillin can be explained by parental health beliefs and demographic variables. Whereas, Barakat et al. (2002) reported that family income, SCD knowledge, and family functioning accounts for 19.6% of the variance seen in adherence. According to Sullivan (2012), irrespective of how the variability of the outcome is obtained, the variability of the outcome should not be too conservative in order to avoid getting a sample size that is too small. Hence, an R^2 value of 0.196, as derived by Barakat et al. (2002) corresponding to a Cohen's f^2 value of 0.244, was used as the effect size.

The G*power software, as described by Faul, Erdfelder, Lang, and Buchner (2007), was used to carry out an a-priori sample size calculation using a statistical power of 0.8, alpha 0.05, a Cohen f^2 effect size of value of 0.244, and number of predictors, six (caregiver knowledge, health beliefs, religiosity, self-efficacy, social support, and vulnerability factors). Power analyses was computed a-priori, with the test family set as *f*-test and the statistical test as linear multiple regression: Fixed model, R^2 deviation from zero. Carrying out this analysis in this way resulted in a minimum sample size of 63 participants. According to Field and Miles (2010) and Field (2013), a sample size of 77 is adequate in a regression model involving six or fewer predictors and a large effect size. However, 100 participants were recruited into this study to make up for any incompletely filled questionnaire that may result to a reduction in sample size.

Recruitment, Participation, and Data Collection Procedures

Before recruiting participants into this study and data collection, permission was received from the medical director of the government-owned hospital and the program director of the Sickle Cell Foundation for the use of their facility as a study site. Ethical approval for the study was received from the Health Research and Ethics Committee of the Lagos State University Teaching Hospital (LASUTH HREC) and from the Institutional Review Board (IRB) of Walden University. A request was also made to the study sites for a health official on duty to assist with the recruitment and data collection. Hence, two health workers (one in each study site) were trained on the need to put participants at ease and not to make any gestures that may influence a participant's response during interviewing. The training involved mock interviews with me and the health workers taking turns as interviewers and respondents. This helped me to look at different anticipated scenarios and standardize the data collection procedure. The health workers also assisted in identifying participants who met the inclusion criteria, recruitment, and data collection.

Prospective participants who met the inclusion criteria were approached in the waiting room, ensured that they understood the English language by asking a few questions in English, provided adequate information about the study, informed that their participation is voluntary, and were given the opportunity to ask questions or any clarifications required. After obtaining voluntary verbal consent, the informed consent form was administered to obtain written consent. Though participants could be allowed to go home with the informed consent form and come back with it at another appointment

date, most participants gave their written consent on site. After obtaining informed consent, data were collected using an interviewer-assisted questionnaire with me or the assigned health professional asking the questions in the questionnaire while marking the answers as provided by the participants and providing any clarifications required by the participants. No identifying information were collected from the participants, but demographic information such as age of caregiver, marital status, family size, number of adults in the house, level of education, religion, employment status, household income, and tribe were collected. Each participant was given a unique identification code that was required for participants who were interested in receiving the results of the study.

Instrumentation

The survey instruments consisted of a combination of items adapted and modified from several validated instruments. Items appropriate to my study were selected from the following instruments:

General information (demographic questionnaire). In addition to caregiver information regarding age of caregiver, marital status, family size, number of adults in the house, level of education, religion, employment status, household income, tribe, religious affiliation, and information on the child were also obtained. Demographic and health information, such as child's SCD type, age of child, caregiver's relationship with child, and monthly/yearly frequency of illness in child, were recorded.

Self-Care Inventory–Sickle Cell (SCI-SC). The SCI-SC is an 18-item parental self-report measure that measures adherence to self-care behaviors in pediatric SCD population (Hilker, Jordan, Jensen, Elkin, & Iyer, 2006). The SCI-SC was developed by

Hilker et al. (2006) in order to identify children at risk of SCD-related complications due to poor adherence to recommended self-care regimen. In using the SCI-SC, parent response on adherence to SCD-self-care resources is rated on a 5-point Likert scale ranging from 1 (*never do it*), 2 (*sometimes follows recommendations but mostly not*), 3 (*follows recommendations about ½ of the time*), 4 (*usually does this as recommended*), to 5 (*always does this as recommended without fail*). Hence, higher scores denote better adherence to SCD self-care management (Hilker et al., 2006). The SCI-SC measures SCD self-care management behaviors such as prescribed medications, avoidance of extreme temperatures (extremely warm or cold), hydration, consumption of a healthy diet, and sleep (Hilker et al., 2006).

Hilker et al. (2006) evaluated the validity and reliability of this measure of adherence by testing this instrument on a population of 99 parents of children with SCD. Hilker et al. evaluated the construct validity of the instrument using correlation with an indirect measure of adherence. Hilker et al. showed that SCI-SC yielded a significantly modest relationship. Item-total correlations and internal consistency were used to assess the reliability of the SCI-SC (Hilker et al. 2006). The researchers found that SCI-SC yielded adequate reliability with item–total correlations by factor ranging from .43 to .77. Hilker et al. also showed that the instrument had a very good internal consistency with a coefficient alpha of .88. Jensen et al. (2005) also evaluated the validity and reliability of the SCI-SC in their study. The study population included parents of children with SCD, aged 6 to 18 years (Jensen et al., 2005). Jensen et al. found significant item-total

correlations ranged between .35 and .77 and a coefficient alpha of .92 for the SCI-SC (Jensen et al., 2005).

The SCI-SC was appropriate as a measure of adherence in this study because it can be used to measure the resources of SCD self-care management as a whole. This instrument was modified in order to identify adherence to individual resources that makes up SCD self-care management, and the scale of the instrument was also changed to a Likert-type continuous scale with values from 0 to 10. Permission to use and to modify this instrument was obtained from one of the developers (Professor Sara Jordan) before use in this study.

Parental health beliefs. Elliott et al. (2001) designed a survey instrument based on the HBM to measure parental health beliefs. This survey instrument contains seven items and is specific to parental beliefs about prophylactic penicillin in pediatric SCD cases. The items in this instrument are rated in a Likert-type scale, ranging from 1 (*least*) to 10 (*most*). Elliott et al. did not provide any evidence for the validity and reliability of this instrument. As the instrument designed by Elliot et al. was specific to prophylaxis penicillin, another instrument was designed (using the format of the instrument by Elliot et al.) The designed instrument contained additional items to account for other SCD self-care management regimen and also used a Likert-type continuous scale.

Sickle Cell Disease Knowledge Test (SCDKT). This is a 10-item test created by Kaslow et al. (2000) for their study. The items in the instrument were designed to measure knowledge about SCD with regards to the genetics of SCD, symptoms, causes of vaso-occlusive episodes, warning signs, and prevention methods. Six of the items in the

instrument are free response items assessing knowledge on SCD characteristics and complications (Hazzard, Celano, Collins, & Markov, 2002). The validity of the items included in the SCDKT were evaluated using the face validity method by a group of professionals and family members of SCD patients serving as judges. Additionally, Kaslow et al. carried out a pilot study on 10 adults to evaluate the reliability of all 10 items in the scale. The researchers reported that the instrument showed a relatively high reliability with a coefficient alpha of .73 (Kaslow et al. 2000). This instrument was appropriate for this study because it can be used to assess parental knowledge of causes and management of SCD. However, only some selected items from this instrument, as appropriate to answer the research question of this study, were used. The instrument is available in the article by Kaslow et al. (2000), but permission to use the instrument was obtained from Professor Nadine Kaslow before use.

Multidimensional Measurement of Religiousness/Spirituality (MMRS). The MMRS was developed by a national working group with the support of the Fetzer Institute and the National Institute of Aging (Fetzer Institute/National Institute on Aging [NIA] Working Group, 1999). The researchers in this working group identified religiosity/spirituality as a construct that was multidimensional and sought to identify the dimensions of religiosity/spirituality associated with health outcomes. The researchers identified 12 dimensions that currently make up the MMRS. These dimensions are daily spiritual experience, meaning, values, beliefs, forgiveness, private religious practices, religious/spiritual coping, religious support, religious/spiritual history, commitment, organizational religiousness, and religious preferences (Fetzer Institute/NIA, 1999).

Several researchers have assessed the validity and reliability of the MMRS as a whole, as well as the validity and reliability of the scales that make up the different dimensions in the MMRS (Harris et al., 2008; Idler et al., 2001; Johnson, Sheets, & Kristeller, 2008; Pargament, Koenig, & Perez, 2000; Pargament, Feuille, & Burdzy, 2011). These studies, among others, evaluated the validity and reliability of the MMRS, its brief version, or part of its dimension on adolescents, college students, elderly patients, and adults of all ages and tribes in the United States.

The most comprehensive evaluation of the brief MMRS was carried out in the 1998 General Social Survey (Idler et al., 2001). Idler et al. (2001) evaluated the validity and reliability of the brief MMRS on 1445 English-speaking adults, aged over 18 years of age. The researchers used component factors analysis and Cronbach's alpha as a measure of the reliability of the brief MMRS. Overall, the different dimensions in the brief MMRS showed an alpha value ranging from .54 to .91. Idler et al. also assessed the validity of the brief MMRS by assessing the content, discriminant, and convergent validity of the instrument. The researchers reported a poor content validity for the measures of religiousness in the brief MMRS (Idler et al. 2001). Idler et al. used correlation matrix as a measure of discriminant validity. The researchers showed that about 80 percent of the significant between-domain correlations were significant with $p < .01$. The researchers also reported that the brief MMRS showed adequate convergent validity, and concluded that the brief MMRS had an adequate reliability and validity (Idler et al. 2001).

For my study, items constituting the dimension of religious/spiritual methods of coping to gain control in the MMRS were evaluated. The items included are items that

assess collaborative religious coping, active religious surrender, passive religious deferral, and pleading for direct intercession as a measure of religious/spiritual methods of coping to gain control. It can be implied from the result of the study by Grossoehme et al. (2013) that religious beliefs and religious coping are related to adherence among parents of children with cystic fibrosis. In this regard, it was necessary to measure the influence of religious coping on adherence; hence, the appropriateness of the MMRS in this study. The MMRS is available free online. However, permission to use the instruments was obtained from the Fetzer Institute and Professor Kenneth I. Pargament of the NIA Working Group.

Sickle Cell Self-Efficacy Scale (SCSES). This instrument is a nine-item instrument developed by Edwards et al. (2000). The items in the SCSES were designed to assess the perceived ability of patients with SCD to manage their disease, as well as take part in everyday functional activities (Edwards et al., 2000). The responses to the items in the SCSES are rated on a 5-point Likert scale ranging from 1 (*not at all sure*) to 5 (*very sure*). The responses to the individual items are summed to obtain a total score, with higher values indicating higher self-efficacy and vice versa (Edwards et al., 2000).

Edwards et al. evaluated the validity and reliability of the SCSES in a population of adults (18 to 73 years) with SCD. The researchers used the internal consistency, Cronbach's alpha as a measure of reliability, and correlation with a set of related constructs to assess the convergent and predictive validity of the scale. The internal consistency showed an alpha value of .89, with all item-total correlation coefficient exceeding .50 (Edwards et al., 2000). The researchers reported that there was a

significant positive correlation, with $p < .01$, between SCSES and other constructs of self-esteem, sense of mastery, and internal health locus of control (Edwards et al., 2000). Clay and Telfair (2007) also evaluated the psychometric properties of the SCSES in a population of adolescents aged 11 to 19 years. The researchers reported a Cronbach's alpha of 0.87 for the nine items in the SCSES. The SCSES is appropriate as a measure of self-efficacy; but the wordings in the instrument were changed to relate to caregivers of children with SCD with permission from Professor Joseph Telfair.

Medical Outcomes Study Social Support Survey (MOS-SSS). Some of the items in the MOS-SSS were used to measure the perceived available social support in this study. The MOS-SSS is a 19-item scale that measures social support in four dimensions emotional-informational support, tangible support, affectionate support, and positive social interaction (Sherbourne & Stewart, 1991). The responses in this instrument ranged from 1 (*none of the time*) to 5 (*all of the time*), with higher scores indicating a higher availability of social support (Sherbourne & Stewart, 1991). Sherbourne and Stewart (1991), staff of the RAND Corporation and the Institute for Health and Aging, developed the 19-item MOS-SSS based on responses gotten from the RAND Medical Outcome Survey involving 2,987 adult patients. The researchers used confirmatory factor analysis to test the overall social support index, used Cronbach's alpha to estimate internal consistency as a measure of reliability, and also used Pearson's product moment correlation between the social support measures and other variables to evaluate the discriminant, construct, and factorial validity of the scales (Sherbourne & Stewart, 1991). Sherborne and Stewart reported a high internal consistency for all the subscales ranging

from .91 to .97, and an alpha of .97 for the total scale. The researchers also found that the MOS-SSS was a valid measure, by showing that all estimated correlations were significant at $p < 0.01$ (Sherbourne & Stewart, 1991).

The MOS-SSS has also been tested in a population of mothers whose children were receiving treatment (Gjesfjeld, Greeno, & Kim, 2008). Gjesfjeld, Greeno, and Kim (2008) reported a high internal validity for the MOS-SSS, and other abridged versions of this instrument with Cronbach's alpha values of .83 and .94 for the 4-item and 12-item MOS-SSS respectively. This instrument was appropriate for this current study as it was used to measure the influence of different dimensions of social support on adherence. The instrument is available free online (http://www.rand.org/content/dam/rand/www/external/health/surveys_tools/mos/mos_socialsupport_survey.pdf). However, permission to use some of the items in this instrument was obtained from the RAND Corporation.

Barriers to Adherence. This was measured using questions developed based on the result of the study by Modi et al. (2009). Modi et al. showed in their study that forgetting, difficulty getting medication, taste and side effect of the medication, among others, were the most reported barriers to adherence among caregivers of children with SCD. The scale was rated in a Likert-type continuous scale from least 0 (*never a problem*) to most 10 (*always a problem*).

To account for the modifications made to some of the validated instruments, and to ensure the validity and reliability of the developed barriers to adherence measure, the validity and reliability of the instruments was assessed. Two medical doctors working

with SCD patients and conversant with research methodologies were consulted to assess if the survey showed construct validity. A pilot study of 10 caregivers of children with SCD was also carried out to identify any potential problems with the survey instrument.

Operationalization of Variables

For the demographic variables, age of caregiver was measured in years and operationalized as a continuous variable. The level of education of the caregiver (*No formal education, Primary education only, some secondary education, secondary school graduate, Tertiary*), and monthly family income (*below 24,999 naira, 24,999-49,999, 50,000-74,999, 75,000 -100,000, above 100,000*) were measured and operationalized on the nominal scale. Marital status was measured and operationalized on a nominal scale (*Single, Married, Separated, Divorced, Widowed*). Other variables such as tribe (*Yoruba, Igbo, Hausa, Others*), relationship of caregiver to child (*Mother, Father, others*), sex of caregiver (*male, female*), and religious affiliation (*Christianity, Islam, Atheist, Traditional religion*) were measured and operationalized on the nominal scale.

The dependent variable, adherence, was measured using some of the items of SCI-SC. Adherence is the degree to which a patient follows the use of a regimen according to the recommendations of the healthcare provider (Dunbar-Jacob, 2004). This variable was measured on the continuous scale, specifically on the interval level of measurement. The instrument contained questions like, "*Does your child drink at least eight glasses of water a day*?" Responses were scored on a Likert-type continuous scale with values from 0 to 10 for each item, and item scores were summed up to obtain a total

score. The adherence measure contained six items, the possible total score ranged from 0 to 60, with higher values signifying better adherence.

The independent variable, parental/caregivers health beliefs referred to the caregiver's perception of the perceived seriousness of SCD in their child/ward, perceived susceptibility to SCD complications, the perceived benefit of the management regimen in preventing complications in their child with SCD, and perceived barriers to complying with the regimen (Elliott, et al., 2001). This variable was measured and operationalized on a continuous scale. An example item in the instrument to measure the HBM indices includes "*How likely is it that you child will get infections?"* Participants were required to rate their response as it corresponds mostly to their extent of agreement from 0 (*least*) to 10 (*most*). Item scores from each construct of the HBM were summed up to get a total score for parental health beliefs.

The parental knowledge variable described the knowledge of parents about methods to prevent SCD complications. An example item in the SCDKT is "*List three symptoms/complications of Sickle Cell Disease*". Correct responses were hand scored (one point for each correct answer), and a total score was obtained by summing across all items, making it possible to measure and operationalize this variable on a continuous scale. The independent variable religiosity/spirituality referred to the belief that human experience is controlled by God which thus influenced the ability to cope with life experiences (Cooper-Effa et al., 2001). The MMRS was used to measure religious coping. An example item in the MMRS is *"I knew that I couldn't handle the situation, so I just expected God to take control".* Responses ranged from 0 (*not at all*) to 10 (*a great*

deal). Religiosity/Spirituality was also measured and operationalized on the continuous scale.

All other independent variables were also measured and operationalized on the continuous scale, with the levels of measurement on the interval scale. The SCSES, which was used to measure caregiver's self-efficacy, contained items like "*How sure are you that you can manage your child's SCD so that you can enjoy doing the things you enjoy doing?"* Responses were measured on a Likert-type continuous scale with scores from 0 (*not at all sure*) to 10 (*very sure*), and items responses were summed to obtain a total score. The social support items in the MOS-SSS included "*How often do you have someone to confide in or talk to about your problems?"* Responses were scored on a Likert-type continuous scale from 0 (*none of the time*) to 10 (*all of the time*). Responses were summed for each item in the MOS-SSS, and a total score was also obtained for total social support available. Barriers to adherence had an example item such as "*Difficulty getting medication*" with response measured on a Likert-type continuous scale ranging from 0 (*never a problem*) to 10 (*always a problem*).

All responses were measured on a Likert-type continuous scale as against a 5-point Likert scale used in the original instrument. This is because it has been posited that it is possible to convert a five point or seven point Likert scale to a continuous variable with a track bar or calibrated line (Allen & Seaman, 2007). Converting Likert scales into a continuous form was reported to add value and variability to data (Allen & Seaman, 2007). In conclusion, all the independent and dependent variables were measured as

continuous variables on the interval scale. Demographic variables contained a combination of interval and nominal scale levels of measurements.

Table 1

Operational Measure Table

Variable Name	Brief Description	No. of items	Response Categories for each item	Type of Variable
Adherence	Measures the degree at which the preventive regimen is used according to the recommendations of the healthcare provider.	6 items	0-10	Continuous
Parental Health beliefs	Measures the HBM constructs: perceived seriousness, perceived susceptibility, perceived benefit, and perceived barriers	16 items	0-10	Continuous
Parental Knowledge	Describes the knowledge of parents about SCD complications and the methods to prevent SCD complications.	4 items	Open ended	Continuous
Religiosity/Spirituality	Measures religious coping	5 items	0-10	Continuous
Self-efficacy	Measures caregivers perceived confidence in taking care of a child with SCD	3 items	0-10	Continuous
Social Support	Measures the presence of perceived financial support, emotional-informational support, and tangible support.	4 items	0-10	Continuous
Barriers to Adherence	Measures the degree at which each barrier to adherence is perceived as a barrier	5 items	0-10	Continuous

Data analysis

The Statistical Package for the Social Sciences (SPSS) version 21 was used to analyze all collected data. The data was examined for missing responses, duplicate entries, and if the minimum and maximum values fall within the expected range for each response using frequency distribution tables. Outliers and values that were out of range were checked using descriptive statistics, scatter plots, frequency distribution tables, or histograms, depending on the level of measurement of the variables. Frequency distribution tables and histograms were also used to assess the distribution of the data. Descriptive statistics was used to describe the participants in terms of demography and other variables.

The research questions and hypotheses for this study are as follows:

RQ1-Quantitative-Descriptive: What is the proportion of parents/caregivers of children with SCD in Lagos, Nigeria, that has adequate knowledge about SCD self-care management?

a. What proportion of parents/caregivers of children with SCD has adequate knowledge of SCD, its complications, and ways of preventing complications?
b. What proportion of parents/caregivers of children with SCD has adequate knowledge of SCD self-care management?

RQ2-Quantitative-Inferential: Is there a relationship between the knowledge of self-care management and compliance with the regimen in children with SCD?

$H_0$2: There is no significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

H_a2: There is a significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

RQ3-Quantitative-Inferential: What are the main predictors or barriers to compliance with SCD self-care management among parents/caregivers of children with SCD?

$H_0$3: No statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

H_a3: Statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

Table 2 shows the different variables measured, the research questions, and the items in the survey measuring these variables.

Table 2

Variables, Research Questions, and Items on Survey

Variable Category	Research Question	Number of Item(s) on Survey
Dependent variable		
Adherence	RQ2, RQ3	Survey Questions 16-21
Independent Variables		
Health Beliefs	RQ3	
• Perceived Seriousness		Survey Questions 22-26
• Perceived Susceptibility		Survey Questions 27-30
• Perceived Benefits		Survey Questions 31-34
• Perceived Burdens		Survey Questions 35-38
Knowledge of SCD self-care management	RQ2	Survey Questions 39-42
Religiosity/Spirituality	RQ3	Survey Questions 43-47
Self-efficacy	RQ3	Survey Questions 48-50
Social Support	RQ3	Survey Questions 51-54
Barriers	RQ3	Survey Questions 55-59
Mediating variables		
Demographic (vulnerability factors) such as total family income, level of education of caregiver, age of caregiver, occupation, gender.	RQ3	Demographic Information, Survey Questions 1-15

The first research question is descriptive in nature; hence, descriptive statistics using frequency tables and charts were used to address this research question. The second research question and hypothesis was addressed using bivariate correlations to evaluate the relationship between knowledge of self-care management which was measured as a continuous variable, and, the continuous dependent variable, adherence. Evaluation of the

data showed that the data was not normally distributed. Hence, Spearman's *rho* test was performed as against the Pearson's *r* test which requires the normal distribution of data.

The third research question and hypothesis was evaluated using regression to determine the relationship between the dependent variable and all predictor variables (independent and mediating variables). Each of the variables measured in this study (predictors or barriers to compliance) were assessed in a separate regression. Multiple regression was used to evaluate the efficacy of the HBM, and other multiple dimension predictors in predicting adherence to SCD self-care management. Multiple regression analysis was also used to assess the main predictors of adherence, as well as the main barriers to adherence. As the assumption of normality and other assumptions for multiple linear regression was not met in the analysis, the dependent variable was recoded into a binary categorical one, and binary logistic regression was conducted instead

The statistical analysis performed for each research question and hypotheses are shown in Table 3.

Table 3

Statistical Analysis for each Research Question and Hypothesis

Research Question	Hypothesis	Variables	Statistical Analysis
RQ1-Quantitative-Descriptive: What is the proportion of parents/caregivers of children with SCD in Lagos, Nigeria, that has adequate knowledge about SCD self-care management?		Knowledge of SCD self-care management	Descriptive statistics
RQ2-Quantitative-Inferential: Is there a relationship between the knowledge of self-care management and compliance with the regimen in children with SCD?	There is a significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD	DV: Adherence IV: Knowledge of SCD self-care management	Bivariate: Correlations between IV and DV. Spearman's *rho* test as the assumption of normality was not met.
RQ3-Quantitative-Inferential: What are the main predictors or barriers to compliance with SCD self-care management among parents/caregivers of children with SCD?	Statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.	DV: Adherence IV: Knowledge of SCD self-care management, Health beliefs, Religiosity/Spirituality, Self-efficacy, Social Support. MV: Demographic factors	Multivariate: DV vs. all IV and MV. Recoding of the DV and binary logistic regression since the assumptions of multiple linear regression was not met

Results are presented in tables as shown in Chapter four. The correlation coefficient, *r*, was used to interpret the relationship between the continuous variables (Field, 2013). The coefficient of determination, R^2, was used to explain the contribution of the independent variable in the amount of variance seen in adherence (Field, 2013). Statistical significance for all the analysis was set at an alpha level of 0.05.

Threats to Validity

Selection bias may have occurred in this study because of the use of the nonprobability based convenience sampling technique. Though, recruiting participants from a children hospital that serve as a reference center for patients with SCD in Lagos, Nigeria, as well as from a sickle cell foundation may have helped reduce this bias. Another source of selection bias in this study was the limitation of participants to those that understand the English language. This may result in the selection of participants of high level of education and high social status, thereby affecting the external validity of this study. To avoid these threats to external validity, the result of this study will not be generalized beyond the population of caregivers of children with SCD that understands the English language, and attending government-owned hospitals in Lagos, Nigeria.

The use of an interviewer in this study may lead to interviewer bias. To minimize interviewer bias, all interviewers (two healthcare professional, one in each study site), were trained on the need to put participants at ease, and not make any gestures that may influence participant's response during interviewing. All instruments used in this study had adequate reliability and validity measures. Hence, the threat to internal validity due to instrumentation was not expected in this study.

Ethical considerations

Permission to use the hospitals and gain access to the participants was obtained from the Lagos State Ministry of Health. Ethical approval for this study was obtained from the health research and ethics committee (HREC) of the Lagos State University Teaching Hospital (approval number LREC/10/06/597). Ethical approval was also

obtained from the Walden University IRB with approval number 11-13-15-0334470. Participants were provided with adequate information about the study, and that no harm will come to them as a result of their participation in the study. Participants were made to know that their participation was voluntary and that they can withdraw their participation at any time during the study with no consequence. All participants recruited into the study gave a written consent by signing an informed consent form. The privacy of the participants was adequately protected by not collecting any identifying information from the participants. Codes were used for sample identification, and data entry was carried out solely by me to further protect participants' data. The collected data is stored in a locked cabinet that is accessible only to me and would be disposed of after five years of this study.

Summary and Transition

A quantitative research method, specifically, a cross-sectional study design was used to explore the factors that influence compliance with SCD self-care management among caregiver's of children with SCD. Using a cross-sectional study design made it possible to collect information on multiple variables in a single study. The target population for this study included caregiver's of children, 5 years and below, with SCD. The inclusion criteria for this target population was that they must be the primary caregiver of a child, 5 years and below, with SCD, they must be attending a government-owned sickle cell clinic in Lagos, Nigeria, and they must understand the English language.

One hundred participants were recruited into this study, with a convenience sampling method used for sampling. Participants attending their regularly scheduled appointments at the pediatric sickle cell clinic were recruited from the waiting area. Participants were briefed about the study, that no harm will come to them as a result of participating in the study, and that their participation is voluntary. All recruited participants gave both verbal and written informed consent. Access to the caregivers was obtained from the Lagos State Ministry of Health, and IRB approval was obtained from the LASUTH HREC and Walden University.

Standardized instruments with adequate validity and reliability measures for data collection were modified and used in this study, and a pilot study was conducted to ensure the adequacy of the modified survey instrument and to assess if further modifications should be done to the survey. Demographic information such as age, education, total family income, among other sociodemographic variables were collected from the participants. Items modified from the SCI-SC was used to assess adherence to SCD self-care management, which is the dependent variable in this study, while items adapted from the HBM, SCDKT, MMRS, SCSES, MOS-SSS, and a developed caregiver's barrier to medication adherence scale was used to collect information on caregiver's health beliefs, caregiver's knowledge of SCD management, religiosity/spirituality, caregiver's self-efficacy, availability of social support, and barriers to adherence respectively.

Statistical analysis was carried out using SPSS 21. Correlations and multiple regression analysis were used to analyze the data in order to answer the research

questions, and results are reported with correlation coefficients and coefficient of determination. The data analysis and result of the study are provided in Chapter 4.

Chapter 4: Results

Introduction

The purpose of this study was to examine the knowledge of self-care management among caregivers of children with SCD in Lagos, Nigeria and to identify the factors that influence compliance with self-care management. Three research questions were postulated for this study. Research Question 1 was descriptive in nature.

RQ1-Quantitative-Descriptive: What is the proportion of parents/caregivers of children with SCD in Lagos, Nigeria, that has adequate knowledge about SCD complications and SCD self-care management?

a. What proportion of parents/caregivers of children with SCD has adequate knowledge of SCD, its complications, and ways of preventing complications?
b. What proportion of parents/caregivers of children with SCD has adequate knowledge of SCD self-care management?

The remaining research questions (Research Questions 2 and 3) were inferential in nature. These research questions and their hypothesis are as follows.

RQ2-Quantitative-Inferential: Is there a relationship between the knowledge of self-care management and compliance with the regimen in children with SCD?

$H_0$2*:* There is no significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

H_a2: There is a significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

RQ3-Quantitative-Inferential: What are the main predictors or barriers to compliance with SCD self-care management among parents/caregivers of children with SCD?

$H_0$3 No statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

H_a3: Statistically significant predictors were detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

In this chapter, I will describe the data collection methods including the timeframe for recruitment, the conduct and findings from the pilot study, the time frame for recruitment and data collection, and the findings from the pilot study. A description of the data analysis using SPSS version 21 including the conduct of statistical assumptions will also be explained. Finally, descriptive statistics will be used to describe the study participants and the demographics of the study participants, and the results of the study per research question are be presented in this chapter.

Pilot Study

To account for the modifications made to the established and validated survey instruments adapted for this study, and because some of the items in the survey were modified to account for caregivers of children with SCD and for peculiar health situations in Nigeria (inclusion of prevention of malaria as part of SCD self-care management in the survey), two medical doctors who were consultant hematologists working with SCD

patients were consulted on whether the questions in the instruments were appropriate to measure the intended variables, especially adherence to self-care management among Nigerian SCD patients. Both doctors validated the survey, giving the survey an adequate face validity. A pilot study of 10 caregivers of children with SCD was then carried out. From the pilot study, I realized that some of the caregiver's dropped out of the university; hence, "*some tertiary education*" was added as one of the options under Question 5 (*What is your Educational Level?*). Also, some of the caregivers mentioned that their child or ward did not fall sick every month. As a result of this, Question 14 was split into two asking the question "*How frequent does your child with SCD fall sick in (a) a month, and (b) a year?"*

Additionally, one of the caregivers encountered during the pilot study just started the clinic as the child was just diagnosed with SCD. This caregiver was not given recommendations on how to take care of her child, and she had no information on what medications to use to help prevent SCD complications in her child. Based on this, an additional exclusion criterion was added to the study. This exclusion criterion helped exclude such caregivers, who had not received any self-care recommendations from the healthcare practitioner, from this study. Apart from these minor corrections, the pilot study did not identify any other potential problems with the survey, and the data collection continued with the modified survey.

D**ata Collection**

Recruitment was done concurrently as data collection. This was because recruitment was done at the waiting area of the study sites. Recruitment and data

collection was done over a 1 1/2 month period from 16th November to 22nd December 2015. Though over 500 caregivers of children with SCD were seen within the data collection frame, about 75% of these had children who were more than 5 years of age; thus, they were excluded from the study. About 45 participants were excluded as they could not read, write, or understand the English language or they were new to the clinic and had not been given any recommendations. In all, the response rate was high as about 95% of the target population meeting the inclusion and exclusion criteria agreed to and took part in the study, making the study population a good representative of the target population.

On average, about 15 surveys were filled out weekly at the government-owned hospital on the 2 clinic days (Wednesday and Friday), as compared to 10 surveys filled out weekly at the SCD group that runs its services daily. At the sickle cell clinic of the government-owned hospital, I was provided a private room for data collection. The nurse assigned to help me with recruitment made a general announcement about my study, the purpose of the study, and that no harm will come to them as a result of their participation in the study. The nurse also announced that participation was voluntary and that the participants were at liberty to take part or otherwise in the study. After taking the vitals of the children, the nurse sent participants who had children 5 years and below to the room I was asked to sit in. From this point, I told the potential participants about the study and the voluntary nature of the study. I again identified that the participants met all the inclusion and exclusion criteria of the study. Then, the data were collected from the participants, after the informed consent form has been signed, with me asking the

participants the question in the survey and ticking the correct response in the survey as answered by the participants. The recruitment method used in the Massey Street Children Hospital helped to increase the participation rate at this study site.

Data entry was done directly into SPSS version 21. Two items under the religious/spiritual methods of coping to gain control had reverse coding. Questions 45 (*Knew that I couldn't handle the situation, so I just expected God to take control*) and Question 47 (*Tried to deal with the situation on my own without God's help*) had response from 0 (not at all) to 10 (a great deal) reverse coded so that the lowest possible score denoted the highest level of spiritual coping. The variable that made up total adherence was coded as a sum of the Survey Questions 16, 17, 18, 19, 20, and 21. The variable that made up parental health beliefs was coded as a sum of Survey Questions 22 to 38. The variable SCD knowledge was coded as the sum of Survey Questions 40, 41, and 42. Spirituality was coded as the sum of Survey Questions 43 to 47, self-efficacy was coded as the sum of Survey Questions 48 to 50, social support was coded as the sum of Survey Questions 51 to 54, and total barriers to adherence was coded as a sum of Survey Questions 55 to 59.

Descriptive and Demographic Statistics

SPSS was used to conduct descriptive statistics. Of the 100 primary caregivers of children with SCD who completed the survey, 60 (60%) were recruited from the government-owned hospital and 40 (40%) from the SCD Foundation. Eighty eight percent of the participants were female and 12% were male. A majority of the caregivers were the parents of the child with SCD with 85% being the mother, 12% being the father,

and 2% being the grandmother of the child with SCD. The age of the caregivers ranged from 20 to 61 years with the participants having a mean age of 34.95. Ninety one percent of the caregivers were married, 3% were single, 5% were separated or divorced, and 1% were widowed. Most of the respondents had medium to high level of education with 40 (40.8%) being high school graduates, 36 (36.7%) having tertiary education, while 2 (2%) dropped out from the university/polytechnic. Fifty six percent of the caregivers knew the type of SCD their child had with 53% being SS and 3% being SC. The remaining 44% did not know the type of SCD their child had. The demographic characteristics of the participants and the disease related variable of their child are presented in Table 4.

Table 4

Demographic Characteristics of Caregivers and Disease-Related Variables of their Child

Characteristics	Percentage (*N*=100)	Mean	*SD*	Range
Caregiver				
Age		34.95	7.43	20-61
Gender				
Male	12			
Female	88			
Relationship with Child				
Mother	85			
Father	12			
Grandmother	2			
Others	1			
Marital Status				
Single	3			
Married	91			
Separated	3			
Divorced	2			
Widowed	1			
Educational level				
No formal education	2			
Primary education only	11			
Some secondary education	7			
Secondary school graduate	40			
Some tertiary	2			
Tertiary	36			
Occupation				
Trader	56			
Artisan	17			
Self-Employed	7			
Employed - White Collar Job	17			
Unemployed	2			
Pensioner	1			

Table Continues

Family Income				
Below 24,999 naira	36			
24,999 - 49,999	22			
50,000 - 74,999	18			
75,000 - 100,000	9			
Above 100,000	9			
Religion				
Christianity	44			
Islam	56			
Tribe				
Yoruba	71			
Igbo	13			
Hausa	3			
Others	13			
Child Age				
one year	13			
two years	20			
three years	19			
Four years	22			
Five years	26			
Gender				
Male	56			
Female	44			
Type of SCD				
SS	53			
SC	3			
Don't Know	44			
Monthly Frequency of illness		0.63	0.89	0 – 5
0	54			
1	32			
2	8			
3	3			
5	1			

Evaluation of the level of adherence to each self-care management resource showed that folic acid supplements was the medication that was most adhered to with a mean adherence value of 9.57 (standard deviation [*SD*] 1.289). This was followed by drinking at least eight glass of water per day with mean adherent value of 9.21 (*SD* 1.604). The mean adherence score and standard deviation of each self-care management regimen is provided in Table 5

Table 5

Adherence Value of each Self-care Management Recommendation

Self-care Management Regimen	Minimum	Maximum	Mean	*SD*	Mean Adherence Rates (%)
Adherence to prophylaxis penicillin	0	10	7.12	4.09	71.2
Adherence to malaria prophylaxis (Proguanil)	0	10	8.82	2.35	88.2
Sleep under insecticide treated nets	0	10	4.08	4.57	40.8
Avoid extreme temperature	0	10	7.79	2.96	77.9
Drank at least 8 glasses of water a day	3	10	9.21	1.60	92.1
Adherence to folic acid supplements	3	10	9.57	1.29	95.7
Total Adherence	24	60	46.52	7.93	77.5

Mean adherence rates were measured as the mean value divided by the maximum possible value for each item then multiplied by 100. Overall or total adherence rates in this study ranged from 40% to 100% with a mean adherence rate of 77.5%. The overall

adherence rates had a median 79.2%, and a 25 and 75 percentile of 66.7% and 84.6% respectively. Sixty one percent of the participants had adherence rates over 75%.

Tests for Statistical Assumptions

In order to establish the adequacy of the hypothesis testing to answer Research Questions 2 and 3, a test of normality of data was carried out. The Shapiro-Wilk's tests and the Kolmogorov-Smirnov tests were used as indicators for normality. Both the Shapiro-Wilk's test and the Kolmogorov-Smirnov test showed statistically significant results ($p < .05$) for the adherence and the SCD knowledge variable; hence, the assumptions of normality distribution were not met and Pearson's Correlation was not suitable to answer Research Question 2. The Spearman's rank correlation was then considered as the statistical test to answer Research Question 2. However, this test requires that the assumption of monotonicity is met. A scatter plot was used to assess the monotonicity of data in the variables total knowledge score and in the items that comprise this variable. Monotonicity of data could be assumed from the scatter plot in all the items and variable tested. Hence, the Spearman's Rank correlation was carried out to answer Research Question 2.

Because the dependent variable did not meet the assumption of normality, the dependent variable was recoded into a binary one. Patel et al. (2010) mentioned that patients who filled their medication 75% of the required time are considered completely adherent. As the total attainable score for adherence in this study was 60, all respondents with total adherence score less than 45 were considered as low adherent, while participants with an adherence score greater than 45 were considered as high adherent. To

check if logistic regression was suitable to analyze the data, the assumptions of linearity of logit and multicollinearity were assessed. To evaluate the assumption of linearity of logit, the continuous independent variables were transformed to their natural log, and binary logistic regression was used to check for interactions between the variables and their natural log. There were no significant interaction effects ($p > .05$); thus, the assumption of linearity of logit was met. A linear regression was used to check the assumption of multicollinearity. The result of this linear regression showed all tolerance values to be greater than 0.1 and all VIF values below 10. The assumption of multicollinearity can, therefore, be assumed in the data (Field, 2013). Binary logistic regression can be used to answer Research Question 3 as all underlying assumption for this statistical test were met.

Research Question 1 Results

What is the proportion of parents/caregivers of children with SCD in Lagos, Nigeria, that has adequate knowledge about SCD complications and SCD self-care management?

One point was given for each correct answer provided in Survey Questions 40 to 42 with the maximum obtainable mark being 10. Knowledge of the complication, causes of complication, and SCD-self-care management were low among the respondents. Total knowledge of SCD was low among respondents with a mean knowledge of 3.82 (*SD* 1.84). In breaking down the results per item that make up the SCD knowledge, the result showed a low knowledge of the things that can cause symptoms/complications in children with SCD with a mean of 0.64 (*SD* 0.86). Knowledge of four things that can be

done to prevent complications of SCD in the respondents had a mean value of 1.51 (*SD* 0.97).

Fifty nine percent of the caregivers could not mention any thing that could cause symptoms/complications in children with SCD. Twenty percent could only mention one correct causes of complication, while 19% could mention two causes of complications. Only 2% of the caregivers could provide three causes of symptoms/complications in children with SCD as asked in Survey Question 41. In assessing the knowledge of self-care management, 14% of the caregivers could not mention any self-care management recommendation, 37% could only mention one self-care management recommendation, and another 37% could mention two recommendations, while 8% could mention three self-care recommendations. Only 4% of the respondents could mention four self-care recommendations as asked in Survey Question 42. In general, only 31% had above average knowledge of SCD complications and self-care management. The remaining 69% had less than average score in the knowledge of SCD. Table 6 shows the mean score of each knowledge element.

Table 6

Mean Scores and Standard Deviation of Total Knowledge and each Knowledge Element

Knowledge item	*N*	Minimum	Maximum	Mean	*SD*
Things that can cause symptoms /complications of SCD	100	0	3	0.64	0.86
Four things that can be done to prevent illness and/or pain crisis	100	0	4	1.51	0.97
Total Knowledge Score	100	1	9	3.82	1.84

Research Question 2 Results

This research question was that "Is there a relationship between the knowledge of self-care management and compliance with the regimen in children with SCD?" The null hypothesis is that there is no significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD, while the alternate hypothesis is that there is a significant relationship between the knowledge of self-care management and compliance with the regimen in children with SCD.

A Spearman's rank-order correlation was conducted to determine the relationship between knowledge of self-care management and total adherence, as well as to determine the relationship between total knowledge and adherence. The result of the analysis showed that there was no significant correlation between knowledge of SCD self-care management and total adherence (r_s = -.049, p = .625). There was also no significant correlation between total knowledge score and adherence (r_s = -.123, p = .223).

Research Question 3 Results

What are the main predictors or barriers to compliance with SCD self-care management among parents/caregivers of children with SCD?

The null hypothesis is that no statistically significant predictors will be detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned while the alternate hypothesis is that statistically significant predictors will be detected as far as the compliance with SCD self-care management among parents/caregivers of children with SCD is concerned.

A binary logistic regression was conducted to assess the relationship between adherence and the independent variables, parental health beliefs, knowledge, social support, self-efficacy, spirituality, and barriers to adherence while controlling for vulnerability factors. Caregiver's educational level, total family income, and monthly frequency of sickness in child were used as the vulnerability/demographic factors. To be able to use binary logistic regression for this analysis, the dependent variable, adherence, was recoded to low adherence and high adherence based on the recommendation by Patel et al. (2010) that patients who filled their medication 75% of the required time could be considered to have complete adherence. As the total attainable score for adherence in this study was 60, all respondents with total adherence score less than 45 where considered as low adherent (Category 1), while participants with adherence score greater than 45 were considered as high adherent (Category 2).

The result of the binary logistic regression to ascertain the effects of parental health beliefs, knowledge, social support, self-efficacy, spirituality, and vulnerability factors on adherence showed that the logistic regression model was statistically significant $\chi^2(15) = 27.588$, $p = .024$. This shows that there is a significant effect for the combined predictors on the dependent variable. From the result, the model explained 49.8% (Nagelkerke R^2) of the variance seen in adherence and correctly classified by 84.4% of the cases. The Hosmer and Lemeshow test also supports the model, χ^2 (8) = 2.869, $p = .942$, which indicates that the model predicted values were not significantly different from the observed. Of the predictor variables included in the model, only three

were statistically significant: knowledge (p = .046), spirituality (p = .031), and barriers (p = .039). Table 7 shows the coefficients in the binary logistic regression model.

Table 7

Coefficients for each Predictor in the Adherence Model

Variable	*b* (SE)	*p*	95% *CI* for Odds ratio		
			Lower	Odds	Upper
Health Belief	-.013 (.013)	.312	.961	.987	1.013
Knowledge	.742 (.372)	.046	1.012	2.100	4.356
Religiosity/Spirituality	-.423 (.196)	.031	.446	.655	.962
Self-Efficacy	-.216 (.141)	.125	.611	.806	1.062
Social Support	.022 (.027)	.418	.969	1.022	1.078
Barriers	-.177 (.086)	.039	.707	.837	.991
Education		.248			
Education 1	49.509 (41299.594)	.999			
Education 2	22.596 (40192.992)	1.000			
Education 3	23.651 (40192.992)	1.000			
Education 4	26.730 (40192.992)	.999			
Total Income		.560			
Total Income (1)	1.451 (1.192)	1.480	.412	4.266	44.150
Total Income (2)	.658 (1.008)	.427	.268	1.931	13.925
Total Income (3)	-1.097 (1.394)	.619	.022	.334	5.129
Total Income (4)	-1.703 (1.949)	.764	.004	.182	8.298
Monthly Frequency of Child's Illness	.856 (.558)	.125	.789	2.355	7.028

Table 7 shows that parental health beliefs did not significantly influence adherence with an odds ratio *OR* = .987 (95% [*CI*] = .961, 1.013), Wald $\chi^2(1) = 1.024$). Apart from parental knowledge, religiosity/spirituality, and caregiver's reported barriers to adherence, not all other variables in the model reached statistical significance. Total parental knowledge influenced adherence in the model with *OR* = 2.10 (95% *CI* = 1.012, 4.356) Wald χ^2 (1) = 3.970, *p* =.046, signifying that those with higher knowledge of the complications of SCD and self-care management were two times more likely to have higher adherence. Spirituality was also a significant predictor of adherence in this model with *OR* = .655 (95% *CI* = .446, .962), Wald χ^2 (1) = 4.659, *p* =.031. The negative logistic coefficient (β value) for spirituality (see Table 7) signifies that increased spirituality was associated with a decrease in adherence. For every unit increase in spirituality, there is a .655 change in adherence. The variable total barriers also reached statistical significance with *OR* = .837 (95% *CI* = .707, .991), Wald χ^2 (1) = 4.253, *p* =.039. This also shows that a one unit increase in total barrier reduces adherence by a factor of .837. Since three statistically significant predictors of adherence were found in the model, the null hypothesis is rejected and it can be concluded that parental knowledge, spirituality, and number of barriers have a statistically significant relationship with adherence.

Effect size and post-hoc power analysis

The Nagelkerke R^2 from the binary logistic regression model is considered a pseudo R^2 value and can be interpreted in the same way as the R^2 value in multiple regression analysis. The R^2 value can be used to estimate Cohen's f^2 which is a measure

of effect size. The Cohen's f^2 can be calculated as R^2 divided by one minus R^2. In this study, the Nagelkerke R^2 value was 0.498. The Cohens f^2 is, therefore, 0.992, which signifies a high effect size. The G*power software was used to carry out a post-hoc analysis to compute achieved power given alpha, sample size, and effect size. The alpha level was set as 0.05, a Cohen f^2 effect size of value of 0.992, number of predictors, seven (caregiver knowledge, health beliefs, religiosity, self-efficacy, social support, total barrier, and vulnerability factors), and sample size was set as 64 since only data from 64 participants was used in the binary logistic regression (due to missing values or incompletely filled survey). Power analyses was computed as post-hoc, with the test family set as f-test, and the statistical test as linear multiple regression: Fixed model, R^2 deviation from zero. The post-hoc analysis showed that a statistical power of 0.999 was attained.

Summary and Transition

The purpose of this study was to determine the proportion of caregiver's of children with SCD that had adequately knowledge of SCD complications and self-care management, as well as to identify the factors that influence adherence. Descriptive statistics was used to describe the demographic characteristics of the respondents in the survey, as well as describe the disease related variables of the child of the caregivers. Spearman's rank correlation analysis was used to determine if there was a significant association between knowledge of SCD complications and SCD self-care management and adherence. Regression analysis was carried out to identify the best predictors of adherence, and the results of the study were organized based on each research question.

Descriptive statistics showed that the proportion of caregivers that had adequate knowledge of SCD self-care management was low, with about 44% not even knowing the type of SCD their child had. Spearman's rank correlation analysis showed that there was no significant relationship between knowledge of SCD self-care management and adherence, and the null hypothesis which stated that there was no significant relationship between knowledge and adherence was not rejected. According to the result of the binary logistic regression, caregiver's knowledge of SCD, religious coping as a measure of religiosity/spirituality, and the number of barriers reported by the caregivers where statistically significant predictors of adherence as seen in the model.

Further discussion and interpretation of the results of the study are presented in the next chapter, Chapter 5. Additionally, the relation of this study with existing body of literature are discussed in the next chapter. Furthermore, the limitations and generalizations of the result of this study, as well as the recommendations for practice and for future research are also discussed in Chapter 5.

Chapter 5: Discussion, Conclusions, and Recommendations

Introduction

The purpose of this quantitative study was to examine if there was an adequate knowledge of self-care management among caregivers of children with SCD in Lagos, Nigeria. The study was also conducted to identify factors that may serve as promoters or barriers to adherence to self-care management among caregivers of children with SCD.

Self-care management is comprised of all actions recommended by the health care practitioner, but carried out at home in the absence of the health care practitioner, and required to improve psychosocial conditions, prevent health complications, and maintain health (Ballas, 2010; Jenerette & Murdaugh, 2008). To identify if there was an adequate knowledge of self-care management among caregivers of children with SCD, as well as to identify the factors influencing caregiver's compliance to SCD self-care management, self-reported information on several variables including demographic information, knowledge of SCD complications and prevention, parental health beliefs, religious coping as a measure of religiosity/spirituality, self-efficacy, social support, number of barriers, and adherence was collected using an interviewer-assisted survey.

The variables were measured on a Likert-type continuous scale from 0 to 10 in order to allow the variables to be measured on a continuous scale, which is the highest level of measurement. Total adherence was calculated as a sum of all the response to the different self-care management recommendations for children 5 years and below. Respondents with ≥75% total adherence were coded as high adherent while respondents

with less than 75% adherent were coded as low adherent as recommended by Patel et al. (2010).

A summary of the key findings of the study as well as the relation of this study with what is available in the literature is provided in this chapter. The limitations of the study and the recommendations for practice and for future research are also described. Finally, this chapter will close with the implications for positive social change and conclusion.

Summary of Key Findings

The total adherence was high in this study with a mean total adherence value of 46.52 (*SD* 7.928). Adherence rates ranged from 40% to 100% with a mean adherence rate of 77.5%. Sixty one percent of the caregivers had adherence rates greater than 75%, which signifies a high level of adherence as reported by Patel et al. (2010). According to the main result of the study, total knowledge was low (mean = 3.82, *SD* 1.844). Knowledge of the causes of complications in children with SCD was also low (mean = 0.64, *SD* 0.859). The knowledge of self-care management as measured in this study was also low with a mean value of 1.51 (*SD* 0.969). Only 2% of the respondents could correctly mention three causes of symptoms/complications in children with SCD, while only 12% of the respondents could mention three or more self-care management recommendations and had adequate knowledge of self-care management. In all, 31% of the caregivers interviewed had above average knowledge of SCD complications and self-care management.

Bivariate correlation of the relationship between knowledge of self-care management and adherence, and total knowledge and adherence, showed no statistical significant correlation between knowledge of SCD self-care management and adherence and total knowledge score and adherence with a Spearman's rho and *p. value* of r_s = -.049, p = .625 and r_s = -.123, p = .223 respectively. However, multivariate analysis identified three variables that influenced adherence to SCD self-care management. These variables (knowledge, spirituality, and total barriers) were the only statistically significant results in the logistic regression with p values of .046, .031, and .039 respectively. The findings from the logistic regression showed that total parental knowledge was a promoter of adherence with *OR* = 2.10 (95% *CI* = 1.012, 4.356) Wald χ^2 (1) = 3.970, while religiosity and total barriers negatively influenced adherence (were barriers to adherence) with *OR* = .655 (95% *CI* = .446, .962), Wald χ^2 (1) = 4.659 and *OR* = .837 (95% *CI* = .707, .991), Wald χ^2 (1) = 4.253 respectively.

Interpretation of the Findings

The results of this study showed that a majority of the participants had a high level of adherence to self-care management recommendations. However, knowledge of SCD complications and prevention of these complications was generally low among the participants. It was expected that participants with higher levels of adherence would have high levels of knowledge of SCD self-care management, but this was not the case in this study. The interpretation of the results of this study in relation to previous studies in peer-reviewed literatures are discussed next.

Adherence to Sickle Cell Disease Self-care Management Recommendations

In this study, 61% of the participants had adherence rates over 75%, with a self-reported overall mean adherence rate of 77.5%. This adherence rate was high compared to the 58.4% adherence rate reported by Patel et al. (2010). Unlike this study (which reported a high level of adherence with a median of 79.2% and the 25 and 75 percentile of 66.7% and 84.6% respectively), Patel et al. reported a median of 59% and a 25 and 75 percentile of 36.6% and 79.6% respectively. One of the limitations in existing literatures is that, apart from the study by Patel et al. where the researchers examined adherence to multiple medications, other researchers only looked at adherence to one medication. Hence, the results of individual regimen that makes up the SCD self-care management are discussed below.

Adherence to prophylaxis penicillin. Adherence to penicillin prophylaxis has continued to be a problem even among the sample of Nigerian caregivers. An adherence rate of 71.2% observed in this study was higher than the adherence rate for prophylaxis penicillin reported in previous studies. This value is still lower than the benchmark for complete adherence, which is 75%. The cost of this medication was one of the major complaints reported by the respondents. Other studies also reported less than optimal adherence rates for penicillin. Elliot et al. (2001) reported 60% adherence to penicillin while others (Bitarães et al., 2008; Patel et al., 2010; Witherspoon & Drotar, 2006) recorded a 48.1%, 54.9%, and 56.7% adherence to prophylactic penicillin, respectively, among caregivers of children with SCD, which are all less than optimal values for

adherence. Similar to the study by Patel et al. (2010), this study also showed that penicillin was the regimen least adhered to among the SCD medications.

Adherence to malaria chemoprophylaxis. There were no studies in the existing body of literature that examined adherence to malaria prophylaxis among children with SCD. This study showed a high adherence to malaria prophylaxis among caregivers of children with SCD with a mean adherence rate of 88.2%. Seventy five percent of the respondents in this study reported 100% adherence to the malaria chemoprophylaxis, proguanil. Kotila, Okesola, and Makanjuola (2007) showed that 69% of adult patients with SCD use proguanil. As the severity of malaria decreases with age, it is expected that children with SCD should have higher adherence to malaria chemoprophylaxis. Eke and Anochie (2003) showed that the use of malaria chemoprophylaxis reduces the number of painful crisis and blood transfusions in children with SCD. With Nigeria being a malaria endemic country, and with the high levels of malaria mortality reported in Nigeria (Malaria Consortium, 2016), it is expected for caregivers to have high adherence to malaria chemoprophylaxis.

Use of insecticide treated nets. Of all the SCD self-care management resources examined in the study, adherence to the use of insecticide treated nets was the lowest with a mean adherence rate of 40.8%. Only 35% of the participants had adherence rates greater than 75%. Kotila et al. (2007) also reported low use of insecticide-treated nets in adult SCD patients with only 10% of the participants in their study sleeping under insecticide treated nets. According to Brousse et al. (2014), SCD patients in malaria endemic African countries should use insecticide-treated nets in combination with

malaria prophylaxis to prevent malaria. However, most caregivers concentrate more on the use of malaria chemoprophylaxis. Some of the reported complaints given by the participants in this study about not adhering to the use of insecticide-treated net were that it causes heat and prevents their child from sleeping well. This is a common barrier considering that Nigeria is a temperate country.

Adherence to folic acid. The adherence rate to folic acid was the highest as in this study. The mean adherence rate for folic acid in this study was 95.7%. This result was somewhat expected as folic acid is the least expensive and easiest to use of all the medications. Patel et al. (2010) also reported folic acid as having the highest adherence rates among all the medications measured in their study. However, Patel et al. reported an adherence rate of 61.3% compared to 95.7% reported in this study.

From this study, caregivers of children with SCD understood the important role of drinking plenty of water on the health of their child. Eighty seven percent of the caregivers adhered fully to the recommendation of drinking up to eight glasses of water per day with a mean adherent value of 92.1%.

Knowledge of Sickle Cell Disease Self-care Management and Adherence

Low levels of parental knowledge of SCD complication and SCD self-care management was reported in this study. In fact, about 44% of the caregivers did not know the type of SCD their child had. Brown, Falusi and Jaudes (1990) mentioned several SCD home-based management recommendations. Although the caregivers in this study were expected to mention only four of these recommendations, 12% of the caregivers could mention three or more self-care recommendations. This indicates an inadequate

knowledge of SCD and SCD self-care management among the caregivers. Caregivers reported low total knowledge of SCD as out of a possible score of 10 for the knowledge test, the mean total knowledge score was 3.82 (38.2%), *SD* = 1.844 and with scores ranging from 1 (10%) to 9 (90%). This result was in contrast to the study by Jensen et al. (2005) who found a moderate knowledge of SCD among parents of children with SCD. Jensen et al. showed that parents had a moderate knowledge of SCD with a mean knowledge score of 71%. There is a similarity in the range of knowledge score reported in this study and that reported by Jensen et al. whereas this study reported that parental knowledge scores ranged from 10% to 90%, Jensen et al. reported a range of 15% to 95% in parental knowledge score.

Bivariate analysis showed no statistically significant relationship between knowledge and adherence (r_s = -.123, p = .223). Although there are contrasting reports on the relationship between knowledge and adherence in the literature, the results of this study corroborates the negative findings by Berkovitch et al. (1998) and Jensen et al. (2005); there was no significant correlation between knowledge and adherence. One possible explanation for this finding, also noticed when interviewing the caregivers, is that the caregivers did not know the names of most of the medications used by their child and why they should use these resources. They, however, were told by the medical practitioner to strictly adhere to these regimens. The reiteration by the medical practitioner about the importance of using the regimens may have influenced the high level of adherence seen in the caregivers. This explanation is supported by Berkovitch et al. who also attributed the high adherence rate seen at baseline in their study to reiteration

of the medical practitioner on the importance of the regimen. Winnick, Lucas, Hartman and Toll (2005) also identified improved communication between the medical practitioner and the family as a strategy to improve adherence.

Factors influencing Adherence to Sickle Cell Disease Self-care Management

Multivariate analysis using binary logistic regression identified three variables that influenced adherence to SCD self-care management. These variables are parental knowledge, religiosity, and total barriers. The results showed that parental knowledge influenced adherence in the model with an *OR* of 2.10, $p = .046$. Those with higher knowledge of the complications of SCD and self-care management were 2 times more likely to have higher adherence. Hence, knowledge was identified as a promoter of adherence in this study. It was interesting that parental knowledge was one of the variables that influenced adherence to SCD self-care management in the multivariate analysis, especially when there was no bivariate relationship between these two variables. Jensen et al. (2005) provided a possible explanation. Because they also did not find any bivariate correlation between knowledge and adherence, Jensen et al. suggested that other variables might influence the variability seen in adherence. I agree with the conclusion of Jensen et al. that "knowledge is not the only, or perhaps most significant, variable affecting adherence, it does not demonstrate the absence of a relationship between knowledge and adherence" (p. 336). This statement supports the result of this study where the absence of a bivariate relationship between knowledge and adherence did not mean an absence of a relationship between these two variables. The significant relationship between knowledge and adherence seen in the multivariate analysis, along

with the significance of other variables, also supports the statement that knowledge is not the most significant variable influencing adherence.

Both religiosity and total barriers also significantly influenced adherence in this study with *OR* = .655, *p* =.031 and *OR* = .837, *p* =.039 respectively. The logistic coefficient (β value) for both religiosity and total barriers were negative. This means that both religiosity and total number of reported barriers are not promoters of adherence, but are barriers to adherence in this study. This study provides the first report of the influence of religiosity on adherence in SCD self-care management. Putting only the items of religiosity in a binary logistic regression model showed that religiosity accounted for 17.8% of the variance seen in adherence, and the item "pleaded with God to make everything work out," which is pleading for direct intercession as a measure of religious/spiritual methods of coping to gain control was the only statistically significant item in religiosity with *OR* = .401, *B* = -.914, Wald χ^2 (1) = 4.604, *p* =.032. Though there were no quantitative studies to serve as comparison for religiosity, I agree with Grossoehme et al. (2013) who reported that low adherent parents conjectured that they trusted God to take care of their child, and they used prayer to change God.

The number of total barriers reported by the parents also served as a statistical significant barrier to adherence. Unlike the study by Modi et al. (2009) and Witherspoon and Drotar (2006) who identified forgetting as the most reported barrier, this study identified cost of medication and difficulty getting medication as the most reported barriers. Apart from the above mentioned variables, all other variables (parental health beliefs, self-efficacy, social support, and vulnerability factors) included in the analysis

did not significantly influence adherence to self-care management. The combination of the variables measured in this study contributed to 49.8% of the variability seen in adherence. This means that other variables that can influence adherence can be identified to improve the model.

The Health Belief Model and Adherence

Parental health beliefs did not influence adherence in this study. Including each item that made up the parental health belief measure in a binary logistic regression model also did not show any significant association between parental health beliefs and adherence. Though this was similar to the result found in the study by Elliott et al. (2001), Elliott et al. found perceived burden of remembering to administer medication as the only significant component of the HBM influencing adherence. Parental health beliefs contributed to 25.6% of the variance in adherence to self-care management. This is close to the 30% variance in adherence to prophylactic penicillin due to parental health beliefs reported by Elliot et al., and in contrast, Witherspoon and Drotar (2006) reported that parental health beliefs accounted for 50% of the variance seen in adherence to prophylactic penicillin.

Limitations of the Study

This study was limited to only care givers of children with SCD attending government-owned sickle cell clinic in Lagos, Nigeria. This study was also limited to caregivers of children, 5 years and below, with SCD who understood the English language. Hence, the results of this study should not be generalized beyond the population of caregivers of children with SCD that understands the English language and

who attend government-owned hospitals in Lagos, Nigeria. There were some issues with missing data in some of the surveys. Though this did not reduce the statistical power of this study as shown in the posthoc power analysis, and missing data analysis showed that the missing values were randomly missing, this missing data may lead to response bias and inadvertently threaten the internal validity of this study. Also, a self-reported measure was used as a measure of adherence. It is possible for some of the caregivers to over-report their adherence, which may in turn affect the internal validity of this study. Other limitations of this study were due to the innate nature of cross-sectional studies. Causal inferences that the identified factors are responsible for the adherence rate seen in the study population cannot be made in this study (Aschengrau & Seage, 2008; Frankfort-Nachmias & Nachmias, 2008).

Recommendations for Future Research and Practice

Considering that the variables measured in this study contributed only 49.8% of the variability in adherence, other measures that can influence adherence can be identified and evaluated in another study. Also, the result of this study still provided contrasting results on the relationship between knowledge and adherence. The relationship between knowledge and SCD among caregivers of children with SCD needs to be further explored with an intervention study, where knowledge and adherence is measured at baseline, and measured again at another point in time after counseling session involving educating parents on SCD self-care management. The intervention study will help to avoid some of the limitations of the cross-sectional study. An additional study can also be carried out to evaluate the role of the different dimensions of

religiosity, such as spiritual experience, meaning, values, beliefs, private religious practices, religious support, religious/spiritual history, commitment, organizational religiousness, and religious preferences, on adherence to SCD self-care management. In addition to this, future research can also be carried out in other states in Nigeria to replicate this research and to provide additional information on the factors influencing adherence to SCD self-care management. It is also recommended for future researches to use a more objective measure of adherence, such as frequency of pharmacy refills or the MEMS, for measuring adherence. Based on the findings of this study, it is recommended that a group counseling section mostly on SCD self-care management and the function and importance of each SCD self-care management regimen be included at every sickle cell clinic.

Social Change Implications

Social change implications, based on the result of this study, can be adapted for both health and clinical practice. Health care practitioners such as health educators, physicians, nurses, will find it useful to introduce counseling which will be used in reiterating the importance of adherence to SCD self-care management in preventing disease complication at every sickle cell clinic appointments. These counseling can be done to all the caregivers of children with SCD at once in the waiting area of the clinic, and should include information on the function of each regimen used in SCD management. Also since religiosity was identified as one of the barriers to adherence, religious leaders can be included in disease management and prevention. As most people revere their religious leaders, getting the message of adherence to self-care management

from them may help improve adherence. Lagos state currently does not have drug mailing services in its pharmacies. Nigeria as a whole also does not have medication mailing services. The introduction of such services in government pharmacies that readily caters for children with SCD may help to prevent the problem of difficulty getting medication and cost of medication.

Conclusion

With SCD being one of the major contributors to infant/childhood mortality in Nigeria (Adeyele & Ofoegbu, 2013), it is important to identify possible ways to reduce childhood mortality due to SCD in Nigeria. I, by this study, offer some of the factors that influence adherence which can be explored for possible intervention in order to improve the life of children with SCD. While most of the caregivers had an adequate level of compliance, effective intervention based on the result of this study can be introduced to improve adherence in those with less than optimal adherence. The information provided in this study represents the first reports of adherence rates of malaria chemoprophylaxis, adherence rates of the use of insecticide treated nets, and the first quantitative reports of the relationship between religiosity and adherence. Recommendations for future researches that can further explain or corroborate the result of the study were made. In conclusion, there is a need to further explore the relationship between knowledge of SCD and adherence, and also include information on SCD self-care management at sickle cell clinics.

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Appendix A: Survey Instrument

Adherence to Self-care Management of Sickle Cell Disease Among Caregivers

Please select one response only to each of the following questions. Please answer all questions on the survey to the best of your knowledge. There are no wrong or right answers. Thank you very much for your time.

DEMOGRAPHIC INFORMATION

1) Gender
    a. Male
    b. Female

2) What is your relationship with the Child with SCD?
    a. Mother
    b. Father
    c. Others (specify) _____

3) What is your Age? _____

4) What is your Marital Status?
    a. Single
    b. Married
    c. Separated
    d. Divorced
    e. Widowed

5) What is your Educational Level?
    a. No formal education
    b. Primary education only
    c. some secondary education
    d. secondary school graduate
    e. Tertiary

6) What is your occupation? _____

7) What is your total family income?
    a. below 24,999 naira
    b. 24,999-49,999
    c. 50,000 -74,999
    d. 75,000 -100,000

e. Above 100,000

8) How many are you in your family? _____

9) How many adults in your family? _____

10) What is your Religion?
    a. Christianity
    b. Islam
    c. Others (Specify) _____

11) What is your Tribe?
    a. Yoruba
    b. Igbo
    c. Hausa
    d. Others

12) How old is your child with SCD? _____

13) What is the gender of your child with SCD?
    a. Male
    b. Female

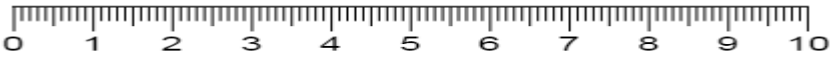
14) How frequent does your child with SCD fall sick in a month? _____

15) How often do you attend scheduled appointments at the pediatric sickle cell clinic, and receive SCD management advice from your healthcare provider
    a. Once a week
    b. Once a Month
    c. Whenever child falls sick
    d. Others (specify) _____

ADHERENCE TO SELF-CARE MANAGEMENT *(This survey has been modified from "Self-care Inventory Sickle Cell" survey from the article "Development of a Screening Instrument of Adherence in Pediatric Sickle Cell Disease" by Hilker, K. A., Jordan, S. S., Jensen, S., Elkin, T. D. and Iyer R.(2006). Children's Health Care, 35(3), 235–246. Copyright © 2006, Lawrence Erlbaum Associates, Inc. Adapted with permission)*

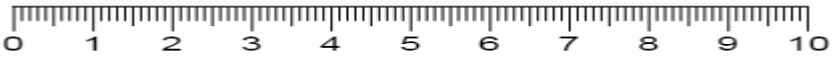
Please provide the most accurate answer to the following questions. On a scale of 0 to 10, score yourself on how often your child/ward with SCD does the following.

16) Take prophylaxis penicillin?



Never do it sometimes always without fail

17) Take malaria prophylaxis (proguanil)?

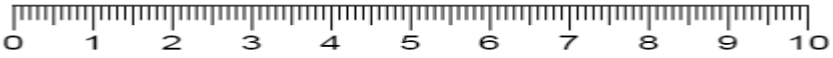


Never do it sometimes always without fail

18) Sleep under insecticide treated bed nets?

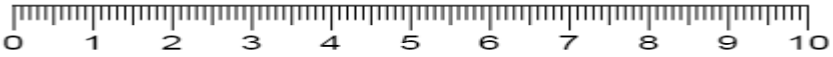
Never do it sometimes always without fail

19) Avoid very warm or cold temperatures?



Never do it sometimes always without fail

20) Drank at least 8 glasses of water a day?



Never do it sometimes always without fail

21) Take folic acid supplements?

Never do it sometimes always without fail

PARENTAL HEALTH BELIEFS

On a scale of 0 to 10, score yourself with the score that corresponds most closely with the extent of your agreement with each question

Seriousness

22) How serious is sickle cell disease?

Not Serious Somewhat Serious Very Serious

23) How serious are infections for your child

Not Serious Somewhat Serious Very Serious

24) How serious is malaria for your child?

Not Serious Somewhat Serious Very Serious

25) How serious is anemia for your child?

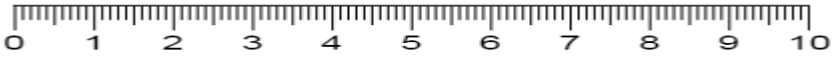
Not Serious Somewhat Serious Very Serious

26) How serious is pain for your child?

Not Serious Somewhat Serious Very Serious

Susceptibility

27) How likely is it that your child will get infections?



Very Unlikely Somewhat Likely Very Likely

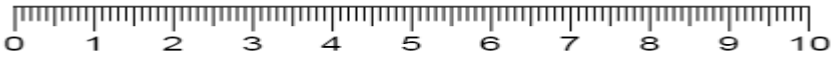
28) How likely is it that your child will get Malaria?

Very Unlikely Somewhat Likely Very Likely

29) How likely is it that your child will get anemia?

Very Unlikely Somewhat Likely Very Likely

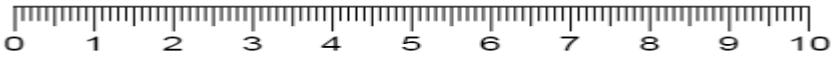
30) How likely is it that your child will get sickle cell pain crisis?



Very Unlikely Somewhat Likely Very Likely

Benefit

31) How well does penicillin help prevent infections for your child?



Not at all Somewhat A great deal

32) How well does proguanil/bed nets help prevents malaria in your child?

Not at all Somewhat A great deal

33) How well does drinking lots of water help prevents pain crisis in your child?

Not at all Somewhat A great deal

34) How well does folic acid helps prevent anemia in your child?

Not at all Somewhat A great deal

Burdens

35) How difficult is it to get the following

Penicillin?

Not at all Somewhat A great deal

Folic acid?

Not at all Somewhat A great deal

Proguanil?

Not at all Somewhat A great deal

Bed nets?

Not at all Somewhat A great deal

Potable water?

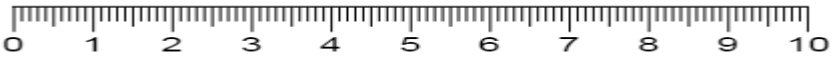
Not at all Somewhat A great deal

36) How difficult is it to administer the following to your child?

Penicillin?

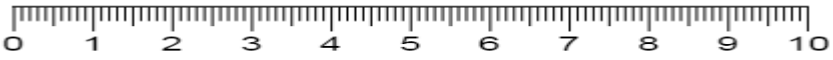
Not at all Somewhat A great deal

Folic acid?



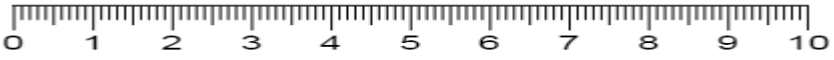
Not at all Somewhat A great deal

Proguanil ?



Not at all Somewhat A great deal

Bed nets?

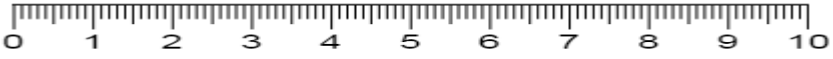


Not at all Somewhat A great deal

Potable water?

Not at all Somewhat A great deal

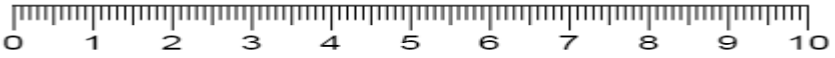
37) How difficult is it to remember to administer the medicine to your child?

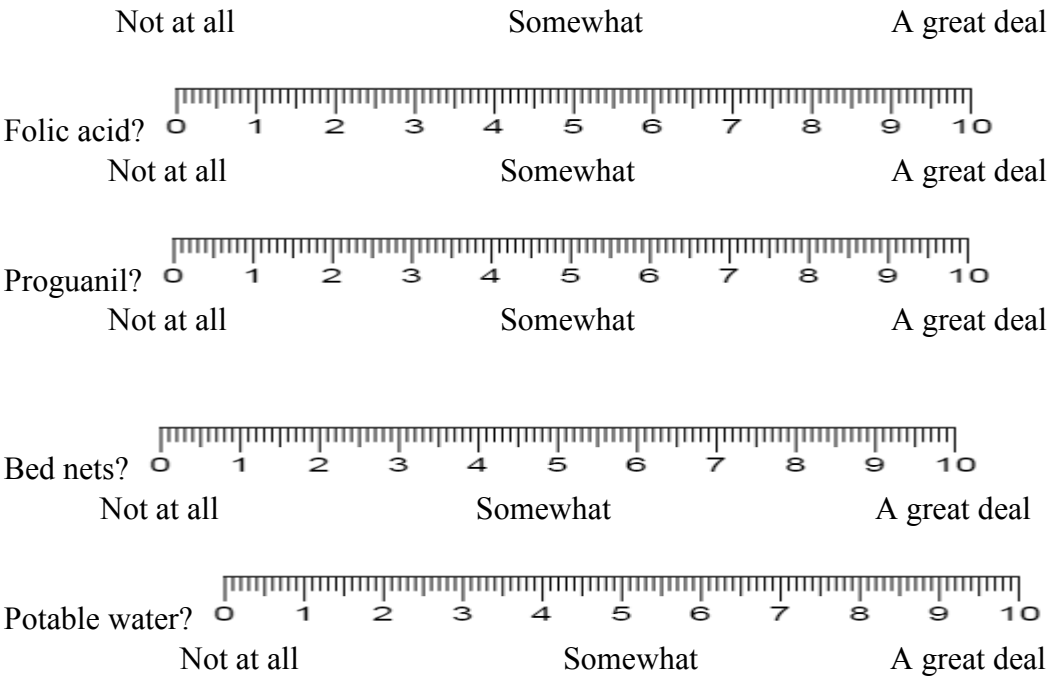


Not at all Somewhat A great deal

38) The following are two costly/expensive

Penicillin?





Sickle Cell Disease Knowledge Test (SCDKT) *(This survey has been adapted with permission from the "Sickle Cell Disease Knowledge Test" from the article "The Efficacy of a Pilot Family Psychoeducational Intervention for Pediatric Sickle Cell Disease (SCD)" by Kaslow, N., Collins, C. H., Rashid, F. L., Baskin, M. L., Griffith, J. R., Hollins, L., & Eckman, J. E. (2000). Families, Systems & Health, Vol. 18, No. 4, 2000 © FSH, Inc.)*

39) What type of Sickle Cell Disease does your child/ward you have? (a) HbSS (b) HbSC (c) HbS-beta thalassemia (d) Do not know

40) List three symptoms/complications of Sickle Cell Disease.
   a.
   b.
   c.

41) List three things that can cause symptoms/complications in children with Sickle Cell Disease.
   a.
   b.
   c.

42) List four things that you can do to keep your child from being ill and/or having pain crises.

a.
b.
c.
d.

RELIGIOUS/SPIRITUAL METHODS OF COPING TO GAIN CONTROL *(This survey has been adapted with permission from the religious coping tool of the "Multidimensional Measurement of Religiousness/Spirituality" survey by the Fetzer Institute/National Institute on Aging Working Group).*

On a scale of 0 to 10, score yourself with the score that corresponds with to what extent you did what the item says.

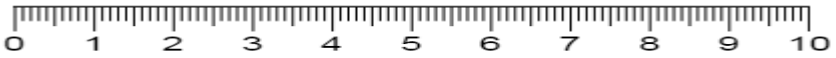
43) Worked together with God to relieve my worries.

Not at all Somewhat A great deal

44) Turned the situation over to God after doing all that I could.

Not at all Somewhat A great deal

45) Knew that I couldn't handle the situation, so I just expected God to take control

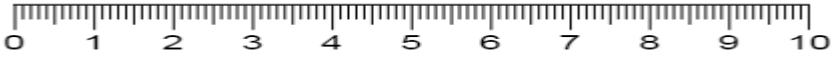


Not at all Somewhat A great deal

46) Pleaded with God to make everything work out.

Not at all Somewhat A great deal

47) Tried to deal with the situation on my own without God's help.

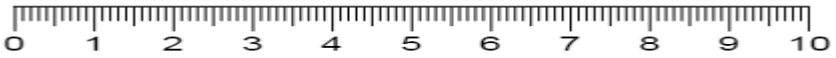


Not at all Somewhat A great deal

SELF-EFFICACY (*(This survey has been adapted with permission from the "Sickle cell Self-efficacy Survey" from the article "Reliability and validity of a self-efficacy instrument specific to sickle cell disease" by Edwards, R., Telfair, J., Cecil, H., & Lenoci, J. (2000). Copyright © 2000 Elsevier Science Ltd.).*

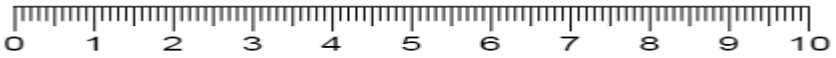
The following questions involve how you deal with having a child with SCD. Score yourself on a scale of 0 to 10.

48) As compared with other people with a child with SCD, how sure are you that you can manage your life from day-to-day?



Not at all sure Somewhat sure Very sure

49) How sure are you that you can manage your child's sickle cell disease so that you can enjoy the things you enjoy doing?



Not at all sure Somewhat sure Very sure

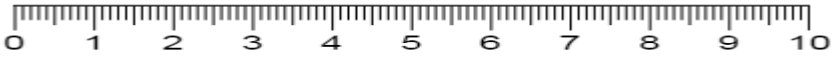
50) How sure are you that you can deal with the frustrations of having a child with SCD?

Not at all sure Somewhat sure Very sure

SOCIAL SUPPORT *(This survey has been adapted with permission from the "Medical Outcomes Study Social Support Survey (MOS-SSS)" copyright the RAND Corporation).*

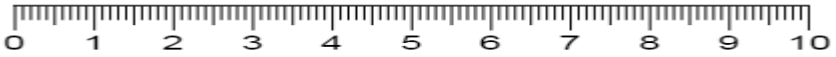
People sometimes look to others for companionship, assistance, or other types of support. How often is each of the following kinds of support available to you if you need it? Circle one number on each line.

51) Someone to confide in or talk to about your problems



None of the time Some of the time All of the time

52) Someone who understands your problems



None of the time Some of the time All of the time

53) Someone to help you financially

None of the time Some of the time All of the time

54) Someone to help you with your child when you are away or ill

None of the time Some of the time All of the time

BARRIERS TO ADHERENCE

Some factors sometimes influence How much of a problem are each of the following in adhering to your child's medications

55) Forgetting.

Never a problem Sometimes always a problem

56) Difficulty getting medication.

Never a problem Sometimes always a problem

57) Time constraint/Busy

Never a problem Sometimes always a problem

58) Cost of medication

Never a problem Sometimes always a problem

59) Taste aversion/side effects

Never a problem Sometimes always a problem

Appendix B: Informed Consent Form

Research Title: Adherence to Self-care Management of Sickle Cell Disease Among Caregivers

You are invited to take part in a research study about parents' knowledge, habits, and challenges related to caring for children with sickle cell disease (SCD). The researcher is inviting people that are the parents of or take care of a child that is not more than five years old, and has sickle cell disease, whether with genotype SS or SC to be in this study. You are invited to take part in this study because you have been identified to have a child with SCD, whether SS or SC, that is five years old or under five years old. This form is part of a process called "informed consent" to allow you to understand this study before deciding whether to take part.

This study is being conducted by a researcher named Muinah Fowora, who is a doctoral student at Walden University.

Background Information

This study is to find out if caregivers (people that take care of children with sickle cell disease [SS/SC]) use the drugs and other health recommendations for children with SCD as told them by the medical doctor. This study will also be used to identify the different factors that influence your using the medications as recommended, as well as, factors that prevent you from using the medications as recommended by your child's medical doctor.

Procedures
If you agree to be in this study, you will be asked to:

- answer some questions on your age, marital status, educational level, income, profession, tribe, your knowledge of how to take care of your child with SCD, and if you follow all guidelines as prescribed by the medical doctor.
- Answering these questions may take up to 30 minutes of your time

Voluntary Nature of the Study

Please, know that your participation is voluntary and you are free to refuse to participate in this study. Everyone will respect your decision of whether or not you choose to be in the study and no one at the XXXXXXXXXXXX will treat you differently if you decide not to be in the study. Also know that if you decide to join the study now, you can still change your mind later, and you can stop at any time if you are not comfortable with the questions.

Risks and Benefits of Being in the Study:
Being in this type of study involves some risk of the minor discomforts that can be encountered in daily life, such as becoming upset. This study is completely anonymous as no information that can be used to identify you will be collected from you. Hence, there is no risk to your confidentiality. There is no physical risk associated with participating in this study. This study will not pose any risk to your health or wellbeing. However, you may be a little inconvenienced as answering the questions may take about 30 minutes of your time.

There are no direct and immediate benefits from participating in this study. However, your participation and the result of this study may help influence policies that may provide welfare packages for children with sickle cell disease, and also influence the introduction of SCD self-care management in genetic counseling which may in the long run help improve the health of children with SCD.

Payment:
There will be no payment as a result of your participation in this study.

Privacy:
Any information you provide will be kept confidential. Any personal information provided will not be used for any purposes outside of this research project. Also, your name or anything else that could identify you will not be used in the study reports. Note that no identifying information will be collected from you and data will be kept secured in locked cabinet and in a pass-word protected computer. Data will be kept for a period of at least 5 years, as required by the university.

Contacts and Questions:
You may ask any questions you have now. Or if you have questions later, you may contact the researcher, Muinah Fowora, via the email address at muinahfowora@waldenu.edu or call XXXXXXXXX. If you want to talk privately about your rights as a participant, you can call XXXXXXXXXX. She is the Lagos State University Teaching Hospital Health Research and Ethics Committee representative who can discuss this with you. Her phone number is XXXXXXXXX. The Lagos State University Teaching Hospital Health Research and Ethics Committee approval number for this study is LREC/10/06/597 and it expires on the 8th of September, 2016.

Obtaining Your Consent

If you feel you understand the study well enough to make a decision about it, please indicate your consent by signing below

Printed Name of Participant

_____

Date of consent _____

Participant's Signature _____

Researcher's Signature _____

Appendix C: Permission to Use the MMRS

Amy Ferguson <aferguson@fetzer.org> Tue, Aug 11, 2015 at 1:25 PM

To: Muinah Fowora <muinah.fowora@waldenu.edu>

Hello,

Many thanks for your query and for news of your dissertation subject. Please accept our approval to reference and use the report for the purposes you've mentioned below. If you are looking for additional information or resources related to the scale, you may want to check the following website, http://www.dsescale.org/, which the BMMRS developer Dr. Lynn Underwood Gordon maintains. She also is available to answer queries from users like yourself.

Best wishes to you in your work and studies,

Amy Ferguson

Fetzer Institute

Kenneth I Pargament <kpargam@bgsu.edu> Wed, Aug 5, 2015 at 2:14 PM

To: Muinah Fowora <muinah.fowora@waldenu.edu>

Dear Mr. Fowora:

I am writing this letter to give you permission to use the religious coping items from the MMRS for your research. Please contact me if you have any questions.

Sincerely,

Kenneth I. Pargament, Ph. D.

Professor Emeritus

Department of Psychology

Bowling Green State University

Bowling Green, Ohio 43403

Appendix D: Permission to use the SCDKT

Kaslow, Nadine <nkaslow@emory.edu> Tue, Aug 4, 2015 at 2:36 PM

To: Muinah Fowora <muinah.fowora@waldenu.edu>

Hi,

You have my permission to use the survey instrument. Best wishes to you with your important research.

Njk

Appendix E: Permission to use the MOS-SSS

Berry, Sandy <berry@rand.org> Thu, Aug 27, 2015 at 4:19 PM

To: Muinah Fowora <muinah.fowora@waldenu.edu>

Dear Muinah

If you have accessed this instrument through the RAND external website www.rand.org under "Surveys and Tools" header you are free to use this as outlined on the web page. Directions for use and attribution are on the web page. If your university requires authorization please count this as authorization for use.

Sandra Berry

Senior Behavioral Scientist

Appendix F: Permission to use the SCSES

Joseph Telfair to you

Hello, you have my permission to use the tool. Please be sure to appropriately cite it and keep me appraised any publication you have from your dissertation. I wish you much success in your career. Thank you for work on behalf of persons with SCD.

Regards

Appendix G: Permission to use the SCISC

THE UNIVERSITY OF
SOUTHERN MISSISSIPPI
DEPARTMENT OF PSYCHOLOGY
XXXXXXXXXXXXXXXXX | www.usm.edu/psy

October 5, 2015

Dear Mr. Fowora:

This letter is intended to grant permission for use the Self Care Inventory-Sickle Cell in your dissertation project. I understand that you intend to alter the scaling to a 10-point Likert-type continuous scale for this particular project. I also am aware that you will revise the item "Taken medication given by the doctor" into two items to address penicillin prophylaxis and well to account for malaria prophylaxis pertinent to your study sample.

You also indicated in your request for permission to use the SCI-SC compliance with the following conditions:

- I will use this survey only for my research study and will not sell or use it with any compensated or curriculum development activities.
- I will include the copyright statement on all copies of the instrument.
- I will print a copy of your permission to use the survey.

Please note that the psychometric properties of the scale have not been evaluated in a Nigerian sample of individuals with sickle cell disease, so take care in examining reliability, and if possible, factor structure and validity, in your sample. I would also appreciate a copy of your findings when your study is completed, so that we can track usage of the scale.

Best of luck with your study and feel free to contact me if you have further questions regarding use of this scale.

Best regards,

Sara S. Jordan, Ph.D.
Associate Professor
Director of Clinical Training