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The Prediction of Behavioral Health Outcomes in Adults with Sickle Cell Disease using the Chronic Disease Outcomes Triad Model

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THE PREDICTION OF BEHAVIORAL HEALTH OUTCOMES IN ADULTS WITH SICKLE CELL DISEASE USING THE CHRONIC DISEASE OUTCOMES TRIAD MODEL

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Lisa Gay Fryar

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To: Dean Ora Strickland
College of Nursing and Health Sciences

This dissertation, written by Lisa Gay Fryar, and entitled The Prediction of Behavioral Health Outcomes in Adults with Sickle Cell Disease using the Chronic Disease Outcomes Triad Model, having been approved in respect to style and intellectual content, is referred to you for judgment.

We have read this dissertation and recommend that it be approved.

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The dissertation of Lisa Gay Fryar is approved.

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Florida International University, 2021

DEDICATION

This dissertation is dedicated to the members of the Fryar family and Hawkins family who have gone before me. It is your courage, strength, support, prayers, dedication, and love of God that guided me with patience and perseverance to complete this project. I humbly salute each of you.

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Lisa Gay Fryar, Ph.D.

ABSTRACT OF THE DISSERTATION

THE PREDICTION OF BEHAVIORAL HEALTH OUTCOMES IN ADULTS WITH SICKLE CELL DISEASE USING THE CHRONIC DISEASE OUTCOMES TRIAD MODEL

by

Lisa Gay Fryar

Florida International University, 2021

Miami, Florida

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The lifelong experience of acute and chronic pain associated with sickle cell disease (SCD) not only has damaging physiological sequelae, but it also can negatively impact affected persons psychologically and socioculturally. These triad of SCD sequelae have an inter-relational and interactional mind-body-social connection that impact the behaviors of adults with SCD. These physiological, psychological, and sociocultural domains comprise the triadic sequelae.

The purpose of this study was to investigate the inter-relational and interactional mind-body-social relationship of the triadic sequelae in SCD as predictors of behavioral health outcomes (i.e., sickle cell fatalism, perceived sickle cell prejudice, and SCD self-efficacy) based on propositions of the Chronic Disease Outcomes Triad (CDOT) Model, which was developed as the basis for this study.

A predictive correlational research design was utilized to study 93 males (n = 29) and females (n = 64) adults with SCD to test the Chronic Disease Outcomes

Triad (CDOT) Model's predictive value of the triadic sequelae related to behavioral health outcomes. Measures completed by participants included Brief Pain Inventory, SCD Symptomatology Scale, Beck's Depression Inventory, Chronic Disease Attitude Scale, Sickle Cell Self-Efficacy Scale, and a Demographic tool. Participants were between the ages of 18 to 75 years old and diagnosed with any genotype of SCD. The majority of participants were diagnosed with genotype *HbSS* (n = 64).

Findings indicated that adults with SCD have pain, other physical and other psychological symptoms, e.g. depression while either working or attending school or not. Participants who reported more physical symptoms were more likely to report more psychological symptoms and depression which interfered with their work and/or school involvement. Participants with more physiological and psychological sequelae, were more likely to have less work and/or school (sociocultural) involvement. Physiological, psychological, and sociocultural (work and/or school involvement) sequelae predicted behavioral health outcomes (fatalism, perceived prejudice, and self-efficacy). Other disease-related and participant-related characteristics were significantly related and predicted triadic sequelae and behavioral health outcomes. All significant relationships were consistent with the hypothesized directions.

It was concluded that adults with SCD have a triad of SCD sequelae that are associated with each other and that predict behavioral health outcomes.

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I. INTRODUCTION

Statement of the Problem

Sickle cell disease (SCD), also known as sickle cell anemia, is one of the most debilitating, unpredictable, and difficult to control chronic diseases. It is a chronic disease experienced by people of color around the world, particularly those with an African heritage. This disorder is a hereditary hematologic disease that can affect almost every organ of the body due to occlusion of small blood vessels with sickled cells that disrupts blood flow and results in acute pain episodes at the time of each sickling event. Multiple sickling events over time, often leads to subsequent chronic pain due to chronic damage to organ tissues that occurs with interrupted blood flow to organs during sickle cell pain “crises,” also referred to as “sickle cell pain episodes”, or “vaso-occlusive crises” (VOCs) (Strickland, Jackson, Gilead, McGuire, & Quarles, 2001; Azar & Wong, 2017; Hulihan, Hassell, Raphael, Smith-Whitley, & Thorpe, 2017). SCD not only has damaging physiological sequelae, but it also can negatively impact those affected by it psychologically and socioculturally based on the severity of the resultant physical symptoms, social stigma, and negative attitudes that often occur as a result of the disease (Strickland et al., 2001). Hence, there are a triad of physiological, psychological, and sociocultural SCD sequelae that impact the behaviors of those affected by the disease (behavioral outcomes) which can ultimately influence the person’s ability to deal with SCD (Strickland et al., 2001; Harris & Wallace, 2012)

Nature, Scope, and Impact of Sickle Cell Disease (SCD)

The number of people diagnosed with SCD in the United States (US) is unknown (Centers for Disease Control and Prevention [CDC], 2016). An estimated 100,000 Americans are impacted by this chronic illness. In the United States, one out of every 365 African Americans and one out of every 16,300 Hispanic-Americans are diagnosed with SCD (CDC, 2016).

Medical advancement has improved the life expectancy of persons living with SCD. During the 1960s and earlier, it was described as a childhood disease because the estimated median survival was 14.3 years with 20% of deaths occurring in the first 2 years of life. One-third of deaths occurred before five-years of age; half between 5 and 30 years; and, only one-sixth occurring after the age of 30 years (Platt et al.,1994; Barriteau & McNaull, 2018). Currently, individuals with the most severe form of SCD, that is Hemoglobin (Hb) variant *Hb SS,* have a median age at death of 42 years for males and 48 years for females; and for *Hb SC* the median age at death is 60 years-old for males and 68 years-old for females (Hassell, 2010; Hulihan et al., 2017).

As with most chronic diseases, there are several physiological, psychological and sociocultural sequelae that are associated with SCD, which profoundly impact the quality of life of persons with the disease (Strickland et al., 2001). The number, nature, and severity of the sequelae in these three domains impact the individual's ability to deal with and cope with their SCD. The greater the number of sequelae, the more debilitating or confining they are likely to be to the

individual. Additionally, the more severe the sequelae, the more difficult it will be for persons affected by SCD to cope with and manage their disease. When individuals are living with SCD, their attitudes and beliefs about the disease may influence their ability to cope with their condition. Negative attitudes and beliefs about their disease such as poor SCD self-efficacy related to management of the disease, fatalistic attitudes towards SCD, and their perceptions of prejudice toward person with SCD can impact their ability to derive positive health outcomes related to the disease (Bayliss, Ellis, & Steiner, 2007; Caird, Camic, & Thomas, 2011; Mathur et al., 2016). Furthermore, obtaining optimal health outcomes relies heavily on medical self-management which is highly influenced by one's perception of their self-efficacy to manage SCD along with many other factors (Bayliss, Ellis, & Steiner, 2007). For example, research indicates that troubled emotional state, low self-efficacy, conflicting personal health beliefs, physical limitations, and the presence of comorbid diseases are barriers that interfere with an individual's ability to adequately self-manage and cope with their disease (Bayliss, Ellis, & Steiner, 2007; Janevic, McLaughlin, Heapy, Thacker, & Piette, 2017). These factors are frequent triadic sequelae (physiological, psychological, and sociocultural consequences) of the illness, that influence attitudes and beliefs an affected person may have about the disease which are important behavioral health outcomes in SCD.

Gaps in Knowledge

Understanding the role that chronic disease triadic sequelae play in helping to mold behavioral health outcomes is important for determining what can be

expected when caring for persons with SCD. Although much attention has been given to studying the physiological, psychological, and sociocultural impact of SCD individually, little attention has been given to studying SCD from the perspective of the additive nature of triadic sequelae and their relationship with behavioral health outcomes. Examples are sickle cell fatalism, perceived sickle cell prejudice, and concepts important to self-care management such as SCD self-efficacy. Persons with SCD experience physiological, psychological, and sociocultural sequelae because of their disease but there is very limited research that has investigated the inter-relational and interactional mind-body-social connections of the disease. Furthermore, research is limited on how these triadic sequelae predict the behavioral health outcomes of persons living with SCD. Therefore, the goal of the current study is to help fill this knowledge gap and increase understanding of how SCD triadic sequelae (physiological, psychological, and sociocultural sequelae) are associated with behavioral health outcomes such as sickle cell fatalism, perceived sickle cell prejudice, and SCD self-efficacy.

Purpose of Study

The purpose of this study is to investigate the inter-relational and interactional relationship of triadic sequelae in SCD as predictors of SCD behavioral health outcomes (i.e., sickle cell fatalism, perceived sickle cell prejudice, and SCD self-efficacy) based on propositions of the Chronic Disease Outcomes Triad (CDOT) Model, which was developed as the basis for this study.

Conceptual Framework: The Chronic Disease Outcomes Triad (CDOT) Model

This study is guided by the Chronic Disease Outcomes Triad (CDOT) Model, a conceptual framework developed by the researcher, which was based on the existing literature regarding chronic diseases and common behavioral health outcomes frequently affiliated with chronic diseases. The sequelae of chronic diseases such as SCD are diverse, and their severity and variation depend on individual and specific disease characteristics. Such characteristics are age of onset of SCD symptoms, and type and nature of disease complications and other pathophysiological aspects of the disease; among which are the degree of development of debilitating effects that interfere with patient functioning and comfort (Harris & Wallace, 2012). In addition to age at diagnosis, the resources that the affected person has available to them influence how well they will be able to care for themselves and their perceptions of their disease (Kristjansdottir et al., 2018). Demographic variables such as age, gender, race, educational level (current school status; how many days missed from school in past school year), socio-economic status as reflected by type of health insurance, current employment or student status, number of reported days missed from work or school in past 12 months, reported genotype of SCD, number of reported vaso-occlusive crises (VOCs) in the past 12 months, as well as number of reported hospitalizations for VOCs, number of months since last hospitalization, and current pain status influence behavioral health outcomes as well.

Triadic Sequelae and Behavioral Health Outcomes

There are three domains of sequelae, which have additive inter-relational and interactional relationships that patients may experience when they have a chronic illness – physiological, psychological, and sociocultural known as the triadic sequelae. According to the Chronic Disease Outcome Triad Model, physiological sequelae are the physical signs and symptoms that result from the disease. Psychological sequelae are mental and emotional signs and symptoms that result from the disease and its complications; and sociocultural sequelae are social and/or role changing consequences of the disease. All sequelae can impact one's behavioral health outcomes of the disease or the actions of individuals living with the disease. Behavioral health outcomes are the attitudes, beliefs, and actions taken by the affected person in relation to the disease. These actions can be positive or negative and ultimately reflect how one is dealing with their chronic illness. For example, sequelae associated with a disease may include pain (physiological sequela) that is felt in a specific area or part of the body that is so severe that it may cause depression (psychological sequela) and prevent the person from working or attending school (sociocultural sequela). The affected person may respond to these sequelae of their illness in such a way that they may believe there is nothing that can be done to improve the course of the disease (fatalism); that others think less of them because they have the disease (perceived disease prejudice); and, they are not capable of managing their symptoms (poor self-efficacy for managing their disease). These are examples of the sequelae and

behavioral health outcomes that will be the focus of this study. See Appendix 1 for CDOT Model.

The Nature and Intensity of Triadic Sequelae

The CDOT Model proposes that the triadic sequelae may range in intensity from mild to severe. Mild sequelae are symptoms and/or conditions that occur infrequently with the disease and/or do not create much discomfort nor debilitating and limiting effects on the individual. Typically, mild sequelae are easily controlled, managed, and treated and they rarely negatively interfere with behavioral health outcomes.

Moderate sequelae are symptoms and/or conditions that occur in association with the disease that are not highly discomforting or may intermittently create discomfort or debilitating and limiting effects on the individual. These symptoms and/or conditions typically are not persistent, may be easily controlled at times, but at other times are difficult to control, manage, and treat. Moderate sequelae can result in negative behavioral health outcomes but not on a frequent basis. Moderate triadic sequelae may be associated with either negative or positive behavioral health outcomes depending on their perceived discomfort. The associated symptoms and conditions of moderate sequelae are typically intermittent and waver in their degree of expression.

Severe sequelae are symptoms and/or conditions that are frequently and persistently present in association with the disease which consistently create discomfort and/or debilitating and limiting effects on the individual. The CDOT

Model considers sequelae to be severe if an individual reports one or more sequela(e) as consistantly present, discomforting, and limiting to their functioning either in the same domain or in multiple domains. These sequelae are perceived by the affected individual as difficult to control, manage, and treat. Severe sequelae consistently interfere with and negatively impact behavioral health outcomes. The CDOT Model propagates that severe triadic sequelae tend to result in multiple negative and more extreme behavioral health outcomes than mild or moderate sequelae.

Application of CDOT Model to Sickle Cell Disease

The nature of triadic sequelae have an inter-relational and interactional connection and influences the behavioral health outcomes related to SCD. The nature and severity of triadic sequelae in SCD are determined by varying factors such as: age at disease diagnosis; pathophysiological aspects of the disease such as hemoglobin genotype; the type and severity of complications the person with SCD has developed as a consequence of the disease; and, debilitating effects resulting from SCD.

The common physiological sequelae in SCD are pain, weakness, yellowing of skin or eyes, vomiting, nausea, heart problems, gall stones, eye trouble, kidney problems, swelling of hands or feet, and shortness of breath (Edwards, Telfair, Cecil, & Lenoci, 2000). Psychological sequelae commonly found in persons with SCD include depression as well as feeling sad, tense or nervous, anxious, short-tempered, worried or concerned, as well as problems coping, sleeping, eating, and

paying attention (Cecilio, dos Santos Periera, dos Santos Pinto, & de Carvalho Torres, 2018; Edwards, Telfair, & Lenoci, 2001). Examples of common sociocultural sequelae of SCD include poor school attendance and unemployment (Coleman, Ellis-Caird, McGowan, & Benjamin, 2016). Sickle cell fatalism (negative health outcome), perceived sickle cell prejudice (negative health outcome), and SCD self-efficacy (positive health outcome) are the behavioral health outcomes that will be evaluated within this study. See Appendix 2 for the SCD Chronic Disease Outcomes Triad Model.

Age at Diagnosis

Individuals with SCD are born with this disease. The earlier in life that children show signs and symptoms of SCD, such as sickle cell pain episodes and swelling of joints, the more likely is the disease to be the most severe form and to result in physiological, psychological and sociocultural sequelae that are more intense, persistent and severe (Archer, Galacteros, & Brugnara, 2015; Azar & Wong, 2017; Jonassaint et al., 2016). Therefore, the earlier age of onset of physiological sequelae, the more likely are sequelae to become severe and occur in multiple domains over time. For patients with SCD, early onset of pain events has been associated with more frequent subsequent sickle-cell related complications in the severe form of the disease, (i.e., hemoglobin *SS* genotype (Dampier et al., 2014).

In infancy sickle cell pain can occur as early as 6 to 12 months of age in the form of dactylitis or swelling of the hands and/or feet, which limits dexterity and

ambulation (Dampier et al. 2014). Painful crises often begin in the first year of life and increase steadily with half of all persons with *SS* and *SC* hemoglobin genotypes reporting a major painful crisis by 4.9 years and 7.1 years of age, respectively (Dampier et al., 2014). With medical advances allowing these individuals to live longer, the symptoms tend to worsen over time which leads to chronic complications (Strickland et al., 2001). Patients with SCD who experience complications at a young age are more likely to experience a progression of severe symptoms or triadic sequelae by adolescence (Hampton, 2014). Also, patients who experience severe triadic sequelae by adolescence or young adulthood will be more likely to exhibit negative attitudes and behavioral health outcomes such as sickle cell fatalism, perceived sickle cell prejudice and SCD self-efficacy (Strickland et al., 2001).

Demographic Characteristics

In addition to age at diagnosis of SCD, one's current age, and gender influence symptoms of SCD and behavioral health outcomes. In the case of SCD, surviving to an older age reflects that the disease is likely to have resulted from a less severe genotype and/or was better managed (Adegbola, 2011; Azar & Wong, 2017). A higher education and socioeconomic status also are personal resources that bode well for those with chronic diseases such as SCD (Jonassaint et al., 2016). Individuals with higher education and socioeconomic status have more resources to take better care for themselves, which leads to better behavioral health outcomes (Jonassaint et al., 2016). Gender is also a factor that is predictor of how well one will take care of themselves, since it is known that females

generally manage chronic diseases more effectively (Bayliss, Ellis, & Steiner, 2007; Caird, Camic, & Thomas, 2011; Arduini, Rodrigues, & Trovo' de Marqui, 2017).

SCD Hemoglobin Genotypes.

The severity of sequelae in SCD varies among the different genotypes. In SCD, pathophysiological genotypes that cause an individual's condition must be considered since they greatly influence the nature and severity of SCD sequelae expressed. The predominant genotypes that cause SCD include Hemoglobin (Hb) *SS,* Hb *SC*, Hb *Sβ*+-thalassemia and Hb Sβo-thalassemia (Saraf et al., 2014). Other rare forms of SCD include hemoglobin *SD* and hemoglobin *SE* (Saraf et al., 2014). Sickle cell hemoglobin S *(SS)* disease is typically associated with more severe sequelae and usually results in more debilitating effects over time (Saraf et al., 2014). Since the Hb *SS* form of SCD tends to result in more severe triadic sequelae, it is typically diagnosed early in life at infancy. Those with less severe forms of the hemoglobin pattern may have symptoms that are so mild and infrequent that they may be diagnosed much later in life (Anderson & Asnani, 2013; Ballas et al., 2012).

SCD Complications.

Complications of SCD include health problems that can occur in almost every organ of the body. Most complications result from sickle cell pain events that interfere with circulation of blood to body organs (Strickland et al., 2001; Booker, Blethyn, Wright, & Greenfield, 2006; Ansari, Moufarrej, Pawlinski, & Gavins, 2017;

Bakshi, Lukombo, Shnol, Belfar, & Krishnamurti, 2017). Organs with very small arterioles are likely to be most negatively impacted as blood flow is decreased and cause ischemia and death of cells that are inadequately oxygenated (Okpala, 1998; Strickland et al., 2001; Ansari et al., 2017; Conran & Belcher, 2018). Some of the more severe complications include stroke, hepatomegaly, loss of kidney function, painful and swelling joints, and leg sores (Barriteau & McNaull, 2018; Conran & Belcher, 2018). Such complications may lead to difficulty ambulating, poor cognition, and limited ability to care for oneself (Frostholm, Hornemann, Ornbol, Fink, & Mehlsen, 2018).

Theoretical Propositions

Based on the CDOT Model and what is known about SCD as a chronic disease, the following propositions will serve as the basis for the study hypotheses.

- Younger age at diagnosis of SCD is associated with more severe hemoglobin genotypes, more complications of SCD, and increased numbers and severity of symptoms and disease consequences across the triadic domains.
- Demographic characteristics, such as older current age, older age at diagnosis, and female gender can influence the behavioral health outcomes of persons with SCD.
- The more severe SCD sequealae will have a negative impact by experiencing more symptoms, and frequent symptoms will be experienced within and across domains, as well as have more negative behavioral health outcomes.
- The more severe and frequent triadic sequelae are, the greater will be the number of hosptializations and emergency room visits and diagnosed complications.

Variables

The key variables used for this study are the triadic sequelae, which are categorized as the physiological, psychological, and sociocultural domains; as well

as behavioral health outcomes. To reduce redundancy and potential confusion within the the research hypotheses and questions of this study, the following definitions related to variable categories will apply:

A. Physiological domain scores refer to SCD physical symptom scores, and pain severity and interference scores.

B. Psychological domain scores refer to SCD psychological symptom scores, and depression scores and depression severity.

C. Sociocultural domain score refers to the participants' employment/school status.

D. Demographic characteristics refer to current age, diagnosed age and gender.

E. Disease-related characteristics refer to presence of SCD complications, and frequency of hospital/emergency room visits.

F. Behavioral health outcomes refer to sickle cell fatalism, perceived sickle cell prejudice scores (negative behavioral health outcomes), and SCD self-efficacy scores (positive behavioral health outcome).

Specific Aims of the Study

Based on the CDOT Model, the triadic sequelae are associated with each other and impact the behavioral health outcomes of persons living with SCD. Given the CDOT Model and the derived SCD-related theoretical propositions stated above, the following research aims and hypotheses will be addressed in this study.

Specific Aim 1: To assess the relationship between the number and severity of SCD sequelae in each of the triadic domains with each other and as predictors of behavioral health outcomes.

Hypotheses:

1. There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores, and a negative relationship of these two domains with the sociocultural domain scores when domain variables are assessed both individually and categorically.
2. There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores with negative behavioral health outcome scores, and a negative relationship with positive behavioral health outcome scores; and, there will be a negative relationship of the sociocultural domain score with negative behavioral health outcome scores and a positive relationship with positive behavioral health outcome scores.
3. There will be a positive relationship between negative behavioral health outcome scores (sickle cell fatalism scores and perceived sickle cell prejudice scores) with each other, and an inverse relationship with positive behavioral health outcome scores (SCD self-efficacy scores) in adults with SCD.

4. The number and severity of triadic domain scores will predict negative and positive behavioral health outcome scores.

Specific Aim 2: To examine the relationship of demographic and disease-related characteristics of adults with SCD as predictors of triadic sequelae and behavioral health outcomes of SCD.

Hypotheses:

1. The more frequently adults with SCD report the presence of diagnosed SCD complications, and hospitalizations/emergency department visits over the past year; the higher will be the number and severity of physiological and psychological domain scores, and the lower will be the sociocultural domain scores.
2. Older age at diagnosis, older current age, and female gender will be positively associated with positive behavioral health outcomes (SCSES) and negatively associated with negative behavioral health outcomes (Fatalism and Perceived Prejudice).

Research Questions:

1. To what degree do the number and severity of physiological and psychological domain scores, and the sociocultural domain score predict diagnosed complications, and hospitalizations/emergency department visits over the past year?

2. To what degree do older age at diagnosis, older current age, and female gender predict behavioral health outcomes, (i.e., SCSES, Fatalism, and Preceived Prejudice)?

Definition of Terms and Operationalization of Variables

Adult with SCD: Any individual who self-reports a medical diagnosis of any genotype SCD or sickle cell anemia (SCA) and who indicates an age of 18 years or older.

Demographic Characteristics: The adult person with SCD's self-reported current characteristics in regards to age, gender, race, educational level, socio-economic status as reflected by type of health insurance, marital status, employment status and/or school status and number of reported days missed from work or school in past 12 months.

Disease Characteristics: The adult person with SCD's self-reported age when SCD was diagnosed, reported HbS genotype, date of last painful episode or VOC, number of hospitalizations and emergency department visits for SCD symptoms in the past 12 months, number of SCD pain episodes or crises in the past 12 months, and SCD complications.

SCD Complications: For the purposes of this study, SCD complications are other reported diagnosed conditions that are attributable to pathological changes resulting from SCD, such as stroke, acute abdomen, kidney failure, leg ulcers, and acute chest syndrome.

Physiological Sequelae: Reported body signs and consequences of SCD, including symptoms, physical discomforts and consequences of SCD at the organ or body system level. This construct will be operationalized by the following:

Pain Frequency: The person's rating of how often pain occurred during the preceding 30 days. The Brief Pain Inventory (BPI) will be used to measure pain frequency in this study (Gjeilo, Stenseth, Wahba, Lydersen, & Klepstad, 2007).

Pain Severity: The person's rating of their perceived discomfort due to pain experienced over the preceding 30 days. The Brief Pain Inventory (BPI) will be used to measure pain severity in this study (Gjeilo et al., 2007).

SCD Physical Symptoms: Eleven body symptoms associated with SCD, which will be rated by the study participant over the preceeding six months using an approach specified by Edwards, Telfair, and Lenoci (2001). The physical symptoms are weakness, yellowing of skin or eyes, vomiting, nausea, pain, heart problems, gall stones, eye trouble, kidney problems, swelling of hands or feet, and shortness of breath (Edwards, Telfair, & Lenoci, 2001).

Psychological Sequelae: Reported signs that one's mind and affective state have been impacted or changed as a consequences of SCD. This construct will be operationalized by the following:

SCD Psychological Symptoms- Eight mental and emotional symptoms associated with SCD, which will be rated by the study participant over the preceding six months using an approach specified by Edwards, Telfair, and Lenoci

(2001). The psychological symptoms include feeling sad, feeling tense or nervous, feeling short-tempered, feeling worried or concerned, problems coping, problems eating, and problems paying attention (Edwards, Telfair, & Lenoci, 2001).

Depression- A mental disorder characterized by sadness, loss of interest or pleasure, feelings of guilt or low self-worth, disturbed sleep or appetite, feelings of tiredness and poor concentration (World Health Organization, 2016). Additionally, loss of energy, feelings of hopelessness, thoughts of suicide, and cognitive and somatic correlates of depression also describes this condition (Aalberg & Marttunen, 2003). This construct will be measured by Beck's Depression Inventory (BDI), a 21-item, self-report rating inventory that measures characteristic attitudes and symptoms of depression (Beck, Ward, Mendelson, Mock, & Ergaugh, 1961).

Sociocultural Sequelae: Reports of how one's social characteristics and attributes have been impacted or changed as a result of having SCD. Within the context of this study, sociocultural sequelae will refer to one's employment status/student status, as well as number of reported days missed from school or work due to SCD related sequelae during the past 12 months.

Behavioral Health Outcomes: How one acts, thinks, or behaves due to disease consequences or sequelae.

Perceived SCD Prejudice- One's beliefs about how others view persons with SCD negatively, unfairly, or prejudicially because of their diagnosis. This

construct will be measured by the Chronic Disease Attitude Scale (Strickland, 2013).

SCD Fatalism- The degree that one believes that one has no control over sickle cell pain (SCP) or other symptoms and negative SCD sequelae, and that those with SCD will have an early death. This concept includes fear, pessimism, predetermination, and death anxiety which may have developed because of a high degree of negative experiences as a result of having SCD. This construct will be measured by the Chronic Disease Attitude Scale (Strickland, 2013).

Sickle Cell Self-Efficacy- Is the person's belief in his or her capacity to perform behaviors necessary to produce desired health outcomes despite current circumstances. This construct will be measured by the Sickle-Cell Self-Efficacy Scale (SCSES) (Edwards et al., 2000).

Significance of Study

This current research study will investigate the relationship of the triadic sequelae on disease-related behavioral health outcomes. It will also assess how the mind-body-social inter-related and interactional triadic sequelae may predict behavioral health outcomes, specifically SCD fatalism, perceived SCD prejudice, and SCD self-efficacy.

Research is limited on how the inter-related and interactional triadic sequelae impact behavioral health outcomes in chronic diseases. The results of this study could benefit individuals with SCD by providing documentation of their perceptions about the prejudice and emotional suffering they perceive as they live

with their condition. Often when persons with SCD encounter the health care system, they are in a critical, vulnerable state. However, health care providers know very little about the patient's suffering beyond the painful episodes and crises which are reflected in their perceptions of discrimination and prejudice of others related to their disease.

Health care providers often know their SCD patients only by their frequent visits to the healthcare facilities and their physical sequelae of the disease. Most health care providers may not be aware or may not consider the other associated sequelae, such as psychological and sociocultural consequences of the disease, and how they affect the individual. Healthcare providers may not adequately focus on the inter-related and interactional connection of triadic sequelae.

This study is also of significance because there are studies that investigate the impact of SCD on the person's quality of life (QOL), health care workers' attitudes and beliefs, and the improvement of medical management (Bhagat, Baviskar, Mudey, & Goyal, 2014; O'Connor et al., 2014; Coleman et al., 2016). There is very limited research that investigates the inter-relational and interactional relationship of this mind-body-social connection representing the whole person. Furthermore, there is little evidence of how these sequelae predict the behavioral health outcomes of persons living with SCD. This study will assess behavioral health outcomes of SCD, specifically sickle cell fatalism and perceived sickle cell prejudice as negative health behaviors and sickle cell self-efficacy as a positive health behavior.

II. REVIEW OF LITERATURE

Introduction

This chapter will provide emperic and theoretical support for a predictive and correlational study that will research the mind-body-social inter-relational and interactional connection of the triadic sequelae. This review and critique of the literature will be structured by the theoretical propositions of the CDOT Model, which was developed based on the literature review which guides this research.

The literature review was conducted via a complete search of electronic databases including the *Cumulative Index to Nursing and Allied Health Literature (CINAHL), PsycInfo*, *Medline,* and *PubMed*. The search included peer-reviewed, research reports in English through 2019. The keywords used for this literature review were: chronic disease, chronic pain, sickle cell disease, physical health consequences and symptoms, mental consequences, emotional consequences, social characteristics and attributes, self-efficacy, medical management, fatalism, depression, pain, unemployment, school attendance, psychological symptoms, and perceived prejudice. Each keyword was combined with the terms sickle cell disease or sickle cell anemia and adults. However fatalism and fatalistic beliefs combined with sickle cell disease and adults were so scant that the term sickle cell disease had to be removed.

The Nature and Impact of Sickle Cell Disease (SCD)

SCD is a severe hemolytic anemia derived from genetic inheritance of the sickle hemoglobin gene (HbS) which causes the hemoglobin molecule to

malfunction. When exposed to low oxygen content, the HbS forms a crystal-like shape which then becomes dehydrated, rigid, and red blood cells become sickled shape (Brewin & Howard, 2017; Clauw, Essex, Pitman, & Jones, 2019). This sickling of red blood cells causes them to break down leading to hemolytic anemia, and to clump together, clogging arterioles and small blood vessels, which result in pain and possible organ damage due to reduced blood flow to affected tissues and organs (Ansara et al., 2017; Hoppe et al., 2011). Pain is the cardinal sequela associated with sickle cell disease which is often intermittent because molecules exposed to oxygen before the sickled molecule's membrane becomes too rigid can revert to a normal shape (Hoppe et al., 2011) .

Etiology of Sickle Cell Disease (SCD)

SCD, an illness with acute and chronic pain patterns, is an inherited autosomal recessive genetic disorder that affects many ethnic minorities throughout the world, especially those that are sub-Saharan African, Western Hemisphere Spanish-speaking (e.g. South America, Caribbean, Central America), Saudi Arabian, East Indian, and Mediterranean (e.g. Turkey, Greece, and Italy) backgrounds (CDC, 2016; Jenerette, Funk, & Murdaugh, 2005). This genetic disorder has different genotypes based on the sickle hemoglobin (*Hb S*) allelle and variant patterns involved (Ashley-Koch, Yang, & Olney, 2000; Saraf et al., 2014). The different genotypes vary in frequencies according to the gene base of the area of the world, mostly in areas of high prevalence of malaria, from which those affected reside (Ashley-Koch, Yang, & Olney, 2000; Saraf et al., 2014).

Sickle hemoglobin is purported to be a genetic adaptive mutation to malaria, which helps protect individuals from the disease in malaria-prone areas of the world (Ashley-Koch, Yang, & Olney, 2000; Archer, Galacteros, & Brugnara, 2015; Ansari et al., 2017). The Hb S variant *Hb SS* (sickle cell anemia) is more prevalent in individuals of African descent and has the highest rates of complications such as vaso-occlusive crises (VOC) and acute chest syndrome (ACS) (Ashley-Koch, Yang, & Olney, 2000; Saraf et al., 2014). The other Hb variants, *Hb S/*β^{o}-thalassemia (also classified as sickle cell anemia), *Hb SC* (sickle hemoglobin C disease), *Hb SD* (sickle hemoglobin D disease), and *Hb SE* (sickle hemoglobin E disease) are more prevalent in Mediterranean, Caribbean, South and Central America, Arab, and East Countries, their most prevalent complications include retinopathy and renal papillary necrosis (Ashley-Koch, Yang, & Olney, 2000; Saraf et al., 2014). Another variant of this disorder is *Hb AS* (sickle cell trait). Sickle cell trait is a carrier state only and does not typically result in sickle cell disease symptoms, but it may be a risk factor for sudden death during extreme physical exertion or in environments that have very low oxygen levels, such as high altitudes (Ashley-Koch, Yang, & Olney, 2000).

Pathophysiology of Sickle Cell Disease (SCD)

As noted previously, SCD is a hemolytic anemia in which the red blood cells become sickled in shape, clump together and break down due to reduced oxygen concentration in the blood, dehydration, or other physiological conditions that cause the fragile S-hemoglobin to hemolyze. Pathophysiological aspects of this

inherited condition includes a group of disorders characteristic of the defective hemoglobin molecule. The defect in the hemoglobin molecule causes the hemoglobin to lose its round, pliable, bioconclave disk shape to become more sickled shape. When the hemoglobin becomes sickled and vaso-occlusion occurs, they result in acute pain crises and anemia, which can lead to a non-functioning spleen, stroke, bone pain, renal damage, gall-stones, and pulmonary hypertension (Paradowski, 2015).

The vaso-occlusive features of SCD are very unique among hemolytic anemias (Steinberg, 1999; Uwaezuoke et al., 2018). SCD causes vascular injuries by occluding small and sometimes large blood vessels leading to vaso-occlusion manifesting with varying degrees of severe, episodic bone pain or abdominal pain (Steinberg, 1999; Uwaezuoke et al., 2018). Vaso-occlusion can occur wherever blood flows (Conran & Belcher, 2018). Therefore, multiple areas are often involved simultaneously and symmetrically in areas of the body such as the chest, back, abdomen, or extremities, which can result in diagnosed complications due to SCD (Steinberg, 1999; Conran & Belcher, 2018).

The most noted SCD complications are: strokes, liver failure, kidney failure, acute abdomen, leg ulcers, and acute chest syndrome. It is also a well-known fact that the complications of SCD are numerous and can affect every organ and/or tissues in the body. Therefore, in addition to pain syndromes, these individuals also experience hematological, neurological, ophthalmological, pulmonary, hepatobiliary, splenic, renal, genitourinary, musculoskeletal, and dermatological

complications (Ballas et al., 2012). However, there is very limited research on the effects of the aforementioned complications, except pain syndromes, and the physiological, psychological, and sociocultural aspects of persons living with SCD. When vaso-occlusion occurs, blood circulation to the affected organ becomes greatly reduced, ischemia occurs which results in pain in the affected organs, and the person experiences painful episodes, often referred to as “sickle cell crises.” These crises may last for days or even weeks (Steinberg, 1999; Conran & Belcher, 2018; Uwaezuoke et al., 2018). Since the vaso-oclusion occurs wherever blood flows, multiple organ damage occurs overtime and increases as the individuals age. The organs most often prominently affected are those with small arteries and capillaries in which sickled cells can lodge and clog (Ansari et al., 2017; Azar & Wong, 2017; Jenerette, Brewer, & Ataga, 2014). These include the brain (which can lead to strokes), the liver, kidneys, lower extremities, and lungs (Azar & Wong, 2017). Injury to bodily organs due to reduced blood flow and ischemia lead to the physiological sequelae in SCD; resulting in debilitating effects that are often complex due to the clinical variability of the disease (Azar & Wong, 2017). Some people with SCD experience symptoms so severe that they require assistance with activities of daily living and miss work and school (Jonassaint et al., 2016).

Common Sequelae in Sickle Cell Disease (SCD)

Pathophysiological aspects of chronic diseases, such as SCD are the functional changes within the body that result from the disease. It is important to know the most common pathophysiological aspects of SCD because they can have a broad impact on multiple systems and bodily organs and have unique

changes on body organs, structures, biochemistry, and symptoms. The pathophysiological changes in SCD can be so extensive that they can disrupt normal daily activities and markedly reduce quality of life (Parmar & Saikia, 2018).

SCD Sequelae in the Physiological Domain

Physiological sequelae of SCD include physical symptoms and physical functional changes within the body resulting from the disease. This includes the biological mechanisms underlying the development of symptoms which may be paired or clustered (Parker, Kimble, Dunbar, & Clark, 2005). These mechanisms may be neurologic, metabolic, molecular, and/or genetic in nature (Parker et al., 2005).

Although acute painful episodes are the most common reason for SCD patients' medical involvement, there are approximately 62 other symptoms or complications of the disease which involve 12 basic systems (O'Connor et al., 2014). The most prevalent SCD sequelae in the physiological domain are primarily due to the disease's negative impact on cardiovascular functioning and key organs that are affected when circulation and cardiovascular processes are hampered. The other complications or symptoms include neurologic, renal, liver, cardiac, opthomalgic, and pulmonary involvement, along with chronic pain (O'Connor et al., 2014). The most noted symptoms include: acute pain episodes or crises, chronic pain, weakness, yellowing of the skin or eyes, vomiting, nausea, heart problems, gall stones, visual problems or "eye trouble," kidney problems, swelling of hands or feet, and shortness of breath (Treadwell, Hassell, Levine, & Keller, 2014).

Hence, physical sequelae and complications of SCD include multi-organ involvement, acute and chronic pain, and neurocognitive deficits that further complicate the affected individual's ability to obtain positive behavioral health outcomes (Treadwell et al., 2014).

Acute and Chronic Pain in SCD: Key Physiological Sequela

Acute and chronic pain are the most prevalent and discomforting physiological sequelae in SCD, which are often difficult to manage by healthcare providers and patients (Anim, Osafo, & Yirdong, 2016; Barriteau & McNaull, 2018; Brown, Weisberg, Balf-Soran, & Sledge, 2015). Acute and chronic pain in SCD requires special attention in the discussion of physiological sequelae of SCD due to their prominence as cardinal symptoms of the disease.

Acute pain is the most frequent initial symptom in SCD and is referred to as "sickle cell pain episodes", "sickle cell crises" or "vaso-occlusive crises" (VOCs) (Jenerette, Funk, & Murdaugh, 2005; Brewin & Howard, 2017). Within the context of this study, the terminology "vaso-occlusive crises" or VOCs will be used to refer to acute sickle cell pain episodes. VOCs are often unpredictable, severe and intermittent and do not require treatment until aflare up is experienced or an impending VOC is suspected to be evolving (Coleman et al., 2016). Evidence shows that the burden of SCD pain typically increases over time from childhood to adolescence, and then to young adulthood as organ damage increases due to repeated VOCs (Hampton, 2014; Barriteau & McNaull, 2018). Many individuals with SCD reported that frequent, unpredictable occurrences of VOCs and other

SCD symptomatology are sometimes so severe that long-term hospitalizations are required (United States Food and Drug Administration (USFDA), 2014).

Chronic pain in patients with SCD often begins to occur after multiple episodes of VOCs have resulted in organ damage due to repeated ischemia of organ tissues. It is usually linked to avascular necrosis of organs, such as bones at the joints, central nervous system sensitization, or neuropathies (Coleman et al., 2016). Although, there is little research on the etiology of chronic pain in patients with SCD, it is believed to appear in patients with frequent acute pain episodes, central nervous system changes, avascular necrosis, and chronic osteomyelitis or ankle ulcers (Ballas et al., 2012). Research revealed that many adult and adolescent patients with SCD who experience recurrent acute pain also experience ongoing persistent or chronic pain (Dampier et al., 2017). Some individuals with SCD experienced a pattern of acute episodic pain superimposed upon chronic pain (Dampier et al., 2017). Others reported living with constant chronic pain even when not experiencing VOCs due to the long-term effects of prior repeated VOCs (USFDA, 2014). Some persons noted pain so severe that they prayed for death (USFDA, 2014).

The Impact of Sickle Cell Pain on Psychological and Sociocultural Domains

Although sickle cell pain is a physiological sequela, it can have a major impact on psychological and sociocultural sequelae and domains. And, pain plays an important part in the holistic functioning of persons with SCD (Coleman et al., 2016).

Research has revealed there is a biopsychosocial impact of sickle cell pain in patients. In the United Kingdom, several studies were conducted in which persons with SCD described how the disease negatively impacted their education, employment, personal independence and relationships (Strickland et al., 2001; Edwards et al., 2005; Coleman et al., 2016). SCD patients also noted a level of suffering and distress that "spills over into all aspects of life" (Thomas & Taylor, 2002; Coleman et al., 2016). Interview studies conducted in both the United Kingdom and Jamaica found that physical, psychological, and social functioning were all adversely affected, and individuals described losses associated with living with SCD as an inability to socialize freely, exercise autonomy, or fully assume desired roles (Anderson & Asnani, 2013; Coleman et al., 2016). Another study revealed the massive effects of depression and anxiety on the health-related quality of life of people living with SCD (Coleman et al., 2016).

SCD Sequelae in the Psychological Domain

SCD sequelae specific to the psychological domain include mental and emotional conditions such as mood or affect, cognitive functioning, and behavior triggered by physical and/or biochemical imbalances that result from chronic diseases (Parker et al., 2005; Umeh & Puddephatt, 2018). This also includes the effects of chronic diseases or their treatment approaches on the mind or mental well-being which are due to medications, the disease process itself, or negative SCD-related social experiences (Parker et al., 2005; Umeh & Puddephatt, 2018). SCD psychological sequelae frequently noted in the literature are depression, anxiety, anger, hostility, and impaired cognitive functioning (Taylor, Stotts,

Humphreys, Treadwell, & Miaskowski, 2013). Depression is the most frequently cited psychological sequela in SCD (Taylor et al., 2013).

Depression and SCD.

Approximately 40% to 50% of adults with SCD experience depression, the most common psychological reaction to chronic pain in this population (Taylor et al., 2013). A study conducted on 50 adults with SCD revealed 44% of participants were mildly to severely depressed as measured by the Beck's Depression Inventory (Taylor et al., 2013). Many cross-sectional and prospective studies conducted over the past three decades overwhelmingly suggest that depression and anxiety are often associated with SCD symptomatology such as pain and disease severity, as well as with healthcare utilization (Holloway, McGill, & Bediako, 2016). According to Smith, Johnston, Rutherford, Hollowell, & Tanabe, (2017), this condition is often associated with a complex social-behavioral health burden with a prevalence of depression and anxiety as high as 27% and 6% respectively.

In general, persons diagnosed with chronic medical illnesses, such as SCD, often report depression stemming from the primary struggles associated with their illness (Morgan et al., 2014). Individuals who reported frequent pain in SCD also had higher somatic symptom burden and were more likely to be depressed or anxious (Dampier et al.,2017). Since individuals with SCD experience significant disease burden, severe painful episodes, and medical complications, these

conditions not only lead to depression and anxiety, but also to frequent emergency room visits (Smith et al., 2017).

Closely associated with depressive symptoms and health-related outcomes is the social devaluation a person experience known as stigma (Holloway, McGill, & Bediako, 2016). Research has shown that the close association of stigma and depressive symptoms has a deleterious impact on health and well-being (Holloway, McGill, & Bediako, 2016). For instance, people who experience high levels of both depression and stigma most likely use fewer health services and are less likely to adhere to treatment regimens which leads to an adverse cycle of depression, stigma, and worse outcomes (Holloway, McGill, & Bediako, 2016).

Other SCD Psychological Sequelae.

There is a positive relationship between pain intensity and negative mood such as depression, unhappiness, anger/hostility, frustration, and worry/anxiousness (Taylor et al., 2013). The psychological state of persons experiencing chronic pain leads an individual to become more vulnerable to a wide range of conditions in addition to depression and anxiety, such as catastrophizing, and poor coping strategies (Taylor et al., 2013).

The psychological phenomenom of pain catastrophizing is often implicated in patients with SCD. Pain catastrophizing is a potent psychological modulator of pain which involves exaggerated negative affective and cognitive appraisals of pain (Sullivan et al., 2001; Mathur et al., 2016). Terms such as rumination, helplessness, and magnification of pain are also associated with patients with SCD

(Quartana, Campbell, & Edwards, 2009; Mathur et al., 2016). According to Mathur and colleagues (2016), catastrophizing was greater when patients were reporting SCD pain compared to other SCD symptomatology. Patients with SCD tended to catastrophize more than patients with other diseases characterized by chronic pain because of a number of disease-related factors, among which are the lifelong and life-threatening nature of SCD, and pain which is severe, episodic and often undertreated (Hollins et al., 2012; Mathur et al., 2016). Pain catastrophizing also is more prevalent in SCD patients that are diagnosed as depressed (Citero et al., 2007; Mathur et al., 2016).

Drug abuse is often noted as a psychological sequela in patients with SCD due to the large amounts of opiates and other potentially addictive drugs that persons with SCD are prescribed to control their pain (Bakshi et al., 2017). The condition of drug abuse is considered because often high doses of opioids are required to adequately treat the pain crises. Many health care professionals often under treat the painful condition because of their negative attitudes towards patients with SCD and their suspicion that these patients have a drug addiction (Institute of Medicine, 2012; Freiermuth et al., 2014; Brennan-Cook, Bonnabeau, Aponte, Augustin, & Tanabe, 2018). Additionally, health care providers often delay pain medication administration and unfairly treat the patients based on negative attitudes and judgments (Institute of Medicine, 2012; Brennan-Cook et al., 2018).

Other research revealed patients with SCD often report feelings of isolation along with feelings of uselessness and helplessness (Booker et al., 2006; Smith et

al., 2017). Past studies have shown that chronic pain is associated with disruptions in memory, however there is very limited research on this phenomenon in adults with SCD (Taylor et al., 2013). Given the neurological consequences of SCD ischemic episodes on the brain, disruptions in memory would be an expected sequela. Cognitive defecits were found in both individuals with overt or silent strokes and individuals without strokes who had SCD (Fields et al., 2016; Jonassaint et al., 2016). Youth with SCD also are at greater risk for anxiety and depression, as well as aggression and delinquency (Smith et al., 2017).

SCD Sequelae in the Sociocultural Domain

Sequelae in the sociocultural domain include demographic, cultural, spiritual, religious and social factors that are affected by a chronic disease, such as social attributes, characteristics, and roles of a person that are altered due to the disease (Parker et al., 2005). These factors reflect the individual's ability to contribute to society, function in social roles, and to be supported by their community and family (Parker et al., 2005; Janevic, McLaughlin, Heapy, Thacker, & Piette, 2017). Among adults with SCD, the ability to work, become educated, and to function in family and community roles can be negatively altered by the disease (Smith et al., 2017).

Research indicates there are limited studies relating to academic achievement in SCD, which is a missed opportunity for improving the lives of individuals with this disease (Jonassaint et al., 2016). Additionally, evidence revealed students with SCD are particularly vulnerable for poor academic

achievement and school dropout (Jonassaint et al., 2016; Fields et al., 2016; Smith et al., 2017). During early childhood, students with SCD are at higher risks for low educational attainment, largely due to multiple disease-related health complications and frequent hospitalizations (Jonassaint et al., 2016). More than one-third of these students will miss at least one-month of classes each school year (Jonassaint et al., 2016). Ten-percent of patients with SCD have overt strokes and 39% have silent cerebral infarcts by the age of 20 years, which can negatively impact school attendance and academic performance (Fields et al., 2016). Moreover, children with a history of stroke or silent cerebral ischemia usually have a cognitive deficit which can require special education (Jonassaint et al., 2016).

In addition to debilitating health problems, individuals with SCD may face social, economic, and academic challenges that create barriers to accessing quality healthcare (Treadwell et al., 2014; Brennan-Cook et al., 2018). The frequency of pain affects individuals with SCD's feelings of self-esteem resulting in problems with maintaining employment and personal relationships (Treadwell et al., 2014). Securing and maintaining employment is extremely challenging and creates financial and social burdens (Treadwell et al., 2014; Brennan-Cook et al., 2018; Smith et al., 2017). Due to lack of understanding of SCD within the community and the workplace, conditions such as drowsiness from pain medications, fatigue from anemia, and multiple absences resulting from pain episodes are not well-received (Treadwell et al., 2014). This can create unstable financial situations that affect self-esteem, and leads to employment discrimination and denial of health insurance (Treadwell et al., 2014). When increased stress-

levels are reported there is often a strong association with pain severity that contributes to absences from work (Smith et al., 2017).

When compared to other chronic illnesses, adults with SCD reported more sociocultural sequelae such as difficulties with employment, finances, child care, and leisure-time activities than adults with diabetes (Treadwell et al., 2014). In comparison to the general healthy population, patients with SCD report higher sociocultural sequelae including higher unemployment rates, less education, lower incomes, and higher rates of being single (Treadwell et al., 2014). Patients with SCD in rural settings have less income, educational attainment, limitations in functioning, and access to health care than those in urban settings (Treadwell et al., 2014).

Issues in the Treatment and Management of SCD

Patients living with SCD experience severe episodic and chronic pain and frequently report poor interpersonal treatment within healthcare settings (Freiermuth et al., 2014; Brennan-Cook et al., 2018; Mathur et al., 2016). Severe and undermanaged SCD pain causes patients to exhibit behaviors that often mimic drug-seeking behaviors (Booker et al., 2006). Healthcare providers often treat these patients as drug-seekers or addicts instead of someone seeking treatment for pain (Brennan-Cook et al., 2018). These behaviors place a strain on patient-provider interaction because providers often misperceive patients as substance abusers (Mathur et al., 2016; Brennan-Cook et al., 2018).

SCD patients often report their pain is poorly managed or staff are not responsive to their complaints (Adegbola, 2011; Brennan-Cook et al., 2018). Moreover, staff perceptions of patient's dependence on pain medications affect pain management and place the patients at risk for "pseudo-addiction" (Adegbola, 2011; Matthie, Hamilton, Wells, & Jenerette, 2015). Pseudo-addiction is described as abnormal behavior developing as a direct consequence of inadequate pain management (Adegbola, 2011). As a result of staff misperceptions, the SCD patient may display behaviors of acting out because of feelings of anger and isolation (Matthie et al., 2015). Due to this negative interaction, providers may perceive the SCD patient as having a behavioral problem, or providers may become frustrated because pain cannot be controlled, and become fearful of inducing drug tolerance or dependence (Adegbola, 2011). Overtime, providers may avoid contact with the SCD patient as an attempt to reduce conflict, and the patient may lose trust in the provider which results in a vicious cycle of inadequate pain management (Adegbola, 2011).

SCD is often mismanaged due to the multiple and often complex nature of the symptomatology associated with it, and it is common for individuals affected by the disease to complain of poor treatment practices from healthcare providers (Booker et al., 2006; Smith et al., 2017). Additionally, lack of adequate knowledge of the disease of healthcare providers who treat people with SCD, as well as family and community, stigmatization, financial distress, and daily disruptions of social interactions result in stressors that have a great impact on behavioral health outcomes (Brown, Weisberg, & Sledge, 2016). The extensive disability and stress

that often ensue from physical complications of SCD often have a negative impact on behavioral health outcomes. These complications can result in poor attitudes and poor self-perceptions of the person affected by SCD and their family members, which often lead to mental/emotional and social sequelae, which may negatively impact the behavior of the individual (Jonassaint et al., 2016).

Unfortunately, individuals with SCD feel their pain experience is not believed and misunderstood, thereby leading to poor treatment practices by healthcare providers (Coleman et al., 2016). It is not unusual for psychosocial aspects of the disease to be poorly assessed and left untreated. Research is limited on psychosocial treatment of pain which limits healthcare providers' understanding of the pain experience and its appropriate care (Coleman et al., 2016). There is a dire need for healthcare providers to explore how adults with SCD experience acute and chronic pain and factors that influence health behaviors, coping, and treatment adherence (Coleman et al., 2016). According to Jonassaint et al. (2016), of the small percentage of SCD patients who frequent the emergency room for treatment of SCD symptoms, it is presumed those patients have a low education level. However, there are no clearly known facts as to what contributes to the high frequency of emergency room visits and hospitalizations among selected patients (Jonassaint et al., 2016). In the general population, there are many studies showing low education increases the risk of hospitalizations (Jonassaint et al., 2016). In a retrospective study conducted on 258 adults with SCD between the ages of 19 years old to 70 years old, data showed patients

without a high school education visited the emergency room three-times as frequently as those with post-secondary education (Jonassaint et al., 2016).

Behavioral Health Outcomes in SCD

Behavioral health outcomes of a chronic disease are how one acts, thinks, or behaves due to disease consequences (Strickland, 2013). Behavioral health outcomes include one's beliefs and attitudes about the disease, one's beliefs about what others think about the disease, one's own perceptions of themselves and others who have the disease, their perceptions of their ability to manage their disease, and their disease self-management behaviors (Strickland, 2013). Positive behavioral health outcomes include behaviors such as positive self-care and coping strategies in relation to the disease (Strickland, 2013). Negative behavioral health outcomes are poor adaptive responses to the disease such as poorer motivation and coping relating to the disease that are typically associated with more severe triadic sequelae (Strickland, 2013).

Fatalism and SCD

In reference to health, fatalism refers to beliefs about causes and controllability of disease: specifically, beliefs that individuals are powerless to influence health or illness because these are controlled by external forces (Ramirez & Carmona, 2018). There are limited studies that focus on fatalism and SCD. According to Strickland et al. (2001), chronic disease fatalism is characterized by the belief that one has no control over the chronic disease and related complications, and that those with specific chronic disease will have an

early death. It is an outcome expectation that includes premature death, fear, pessimism, predetermination, and death anxiety (Strickland et al., 2001). Chronic disease fatalism is a matter of degree, i.e., it can occur along a continuum of very little or to an extreme degree (Strickland et al., 2001). Those individuals high in chronic disease fatalism are likely to have developed this attitude because of a high degree of suffering from the disease and/or due to negative attitudes and beliefs about the chronic disease and its outcomes by those with whom they have contact, such as family members, healthcare providers, friends and other acquaintances (Strickland et al., 2001). A large amount of chronic disease fatalism is likely to lead to negative emotional responses such as depression, anger, anxiety, and hopelessness (Strickland et al., 2001). Persons living with sickle cell disease experience this also. Individuals with sickle cell fatalistic attitudes tend to have an external locus of control; they buy into the idea that sickle cell health outcomes are not controllable; and, they are less motivated to believe that personal actions will facilitate positive sickle cell health outcomes such as a reduction in the frequency and severity of sickle cell pain episodes or a decrease in symptoms and complications (Strickland et al., 2001). Additionally, high sickle cell fatalism is likely to have developed because of perceptions about SCD in persons they frequently encounter such as family members, health care providers, friends, and other acquaintances (Strickland et al., 2001).

Along with psychosocial and physical health issues, adults with SCD experience spiritual issues such as hopelessness, negative self-talk, and fatalism (Adegbola, 2011). Often the fatalism experienced by persons with SCD amounts

to a feeling that there is little that can be done to change the course of the disease, which results in minimal actions for self-care due to feeling of despair (Adegbola, 2011). Similar to research on SCD, studies on cancer fatalism also described patient beliefs that developing cancer is beyond one's control and that death is inevitable upon diagnosis (Ramirez & Carmona, 2018). Furthermore, persons with cancer who held fatalistic beliefs tended to have lower rates of cancer screening, were less likely to engage in healthy behaviors such as regular exercise and a healthy diet, and had lower levels of cancer prevention knowledge (Ramirez & Carmona, 2018). In addition, research reveals African-Americans, Latinos, and immigrant populations are more likely to possess fatalistic beliefs when confronted with a chronic disease than persons from other cultures (Ramirez & Carmona, 2018).

Perceived Prejudice and SCD

Perceived chronic disease prejudice is the belief that others view persons with a specified chronic disease negatively, unfairly or in a biased way because they have the disease (Carlisle, 2015). Perceived discrimination, also referred to as perceived prejudice, has been shown to occur in relation to several chronic illnesses and conditions such as hypertension and poor birth outcomes (Carlisle, 2015). Unfortunately, research is limited on the impact of perceived prejudice and chronic health conditions (Carlisle, 2015). However, it is suggested that studies explaining the link between perceived prejudice and health may reveal discrimination is a chronic stressor for persons with some health conditions which may directly lead to negative health outcomes (Carlisle, 2015; Haywood et al.,

2014). Furthermore, perceived prejudice as a chronic stressor in relation to a disease is thought to produce equivalent negative outcomes as other chronic stressors in an individual's life (Carlisle, 2015; Haywood et al., 2014; Mathur et al., 2016). The prevalence of chronic stress related to perceived prejudice about a disease may result in even greater susceptibility to negative chronic diseases outcomes and illnesses and chronic pain conditions (Carlisle, 2015). In social settings, conditions such as low socioeconomic status and perceived prejudice may serve as fundamental causes of the psychosocial responses that lead to chronic stress conditions (Carlisle, 2015).

Racial and ethnic disparities in relation to the burden of pain and quality of pain treatment in SCD are a significant public health problem in the US (Haywood et al., 2014). Research revealed many healthcare providers have exhibited negative attitudes about SCD patients that create significant barriers to the delivery of appropriate pain management (Haywood et al., 2014). In a study conducted on 291 African-Americans that reported having SCD, it was found these patients reported a higher level of race-based discrimination from healthcare providers than other groups of African-American patients (Haywood et al., 2014). The study revealed the sample reported a greater level of discrimination from healthcare providers on the basis of their SCD diagnosis rather than their race or ethnicity (Haywood et al., 2014). This finding indicated a greater association with disease-based discrimination versus race-based discrimination (Haywood et al., 2014). This study further indicated that both disease-based and race-based discrimination were found to be independently associated with SCD patients having difficulty

convincing healthcare providers about the presence and treatment of pain (Haywood et al., 2014). Disease-based discrimination was also found to be independently associated with greater emergency room utilization for SCD pain (Haywood et al., 2014). Patients' perceived that when there was a dispute between them and healthcare workers regarding their pain, the basis of the difficulty was attributed to both their race/ethnic status and their status of having SCD; with the latter receiving greater attribution (Haywood et al., 2014). Research has revealed discrimination in healthcare settings may affect multiple health outcomes including pain management, clinical pain, and pain sensitivity in SCD patients, therefore suggesting that negative interpersonal relationships such as discrimination play a pivotal role in inadequate treatment and pain management in SCD (Mathur et al., 2016). As a result of negative interpersonal experiences, many persons with SCD frequently delay seeking treatment, try to manage VOCs at home, or self-discharge from hospitals earlier than they should (Mathur et al., 2016).

Within the context of the Strickland et al. study (2001), perceived sickle cell disease prejudice was identified as a negative behavioral health outcome, and it was revealed that persons who had attitudes characterized by perceived sickle cell prejudice were preoccupied with feelings of being thought of negatively or of being discriminated against because of their diagnosis of SCD (Strickland et al., 2001). Feelings of disenfranchisement or of being left out of activities and special events by others because of their disease were common (Strickland et al., 2001; O'Connor et al., 2014; Bakshi et al., 2017). Persons with SCD who perceived sickle cell prejudice felt that having SCD kept them at a certain distance in their relationships

because they were stereotyped (Strickland et al., 2001; O'Connor et al., 2014). Among the common stereotypes were that persons with SCD could not "hold down a job," that they were not capable of being physically active, were "sickly," that they should not expect to be involved in successful dating relationships, or that they were "drug addicts" due to the need to control sickle cell pain with opiates (Strickland et al., 2001; O'Connor et al., 2014; Bakshi et al., 2017). Several persons with SCD noted that common sources of these perceptions were friends, family members, potential employers, staff members at hospitals, nurses and doctors (Strickland et al., 2001). Perceived sickle cell prejudice is an attitude that results from perceptions by persons with SCD concerning interactions with people who are aware of their diagnosis (Strickland et al., 2001). Ultimately, it reflects how accepted those with SCD feel they are by others in their environment (Strickland et al., 2001). Persons with SCD who are highly symptomatic are likely to be more sensitive to or perceptive of sickle cell prejudice (Strickland et al., 2001; O'Connor et al., 2014). Those who are preoccupied with sickle cell prejudice also are less likely to be motivated to pursue a full productive life characterized by self-sufficiency, employment, career planning, and intimate personal relationships (Strickland et al., 2001). These persons also are more likely to be characterized by negative emotions such as depression, anxiety, anger, and feelings of hopelessness (Strickland et al., 2001).

Self-efficacy and SCD

SCD is best managed by promoting holistic health and comprehensive self-care strategies that decrease hospitalizations, increase psychosocial functioning,

improve quality of life (QOL), and prevent premature deaths (Adegbola, 2011). Self-efficacy, social support, sociodemographics, and self-care have been described as critical concepts associated with the management of SCD and other chronic diseases (Matthie, Jenerette, & McMillan, 2015). When a person with SCD demonstrates self-efficacy by taking charge of life situations and demonstrating a strong expectation of being successful with the outcomes, coping with this illness is better (Adegbola, 2011). In chronic diseases, higher levels of self-efficacy were linked to decreased pain severity, fewer self-reported symptoms, increased use of adaptive coping mechanisms, and increased adherence to medical regimens (Matthie, Jenerette, & McMillan, 2015). Alternatively, lower levels of self-efficacy were correlated with more symptoms, higher pain severity, and frequent physician visits (Matthie, Jenerette, & McMillan, 2015).

According to Strickland et al. (2001), self-efficacy in SCD is an individual's self appraisal of their ability to engage in daily functional activities despite having SCD. Individuals form self-efficacy beliefs by interpreting information from four sources: mastery experience, vicarious experience, social persuasion, and somatic and emotional states (Redmond & Slaugenhoup, 2016). Mastery experience is interpreting results of one's previous performance. Vicarious experience is interpreting results by observing others (Redmond & Slaugenhoup, 2016). Additionally, individuals also create and develop self-efficacy beliefs as a result of social persuasion received from others and as a response to somatic and emotional states such as anxiety, stress, arousal, and mood states (Redmond & Slaugenhoup, 2016).

Summary

This chapter provided available empiric and theoretical support for this predictive and correlational study that will research the mind-body-social inter-relational and interactional connection of the triadic sequelae. Furthermore, this research will consider how the triadic sequelae may predict behavioral health outcomes of adults with SCD. The major components of the CDOT Model were examined in relation to the research literature. Pathophysiological aspects of SCD, and debilitating effects of SCD were reviewed as well as common sequelae in each triadic domain. Common sequelae of adults with SCD were identified for: physiological, psychological and sociocultural domains. Literature on behavioral health outcomes, that is, sickle cell fatalism, perceived sickle cell prejudice, and SCD self-efficacy was also reviewed to serve as a basis for developing the study's research hypotheses and approaches.

III. RESEARCH DESIGN AND METHODOLOGY

Research Design

This quantitative study used a predictive correlational research design to test the Chronic Disease Outcomes Triad (CDOT) Model to investigate the predictive nature of triadic disease sequelae on behavioral health outcomes in sickle cell disease (SCD). In this study, the triadic sequelae included physiological (pain, other physical SCD symptoms), psychological (depression and other psychological SCD symptoms), and sociocultural variables (employment status and/or school attendance) of adults with SCD as revealed in the literature as some of the more common sequelae experienced by this population.

Predictor variables.

The primary predictor variables were the common triadic sequelae identified from the literature common for patients with SCD. The triadic sequelae included the identified disease sequelae within the physiological, psychological, and sociocultural domains.

Criterion variables.

The primary criterion variables for this study were both positive and negative behavioral health outcomes that included sickle cell self-efficacy, sickle cell fatalism, and perceived sickle cell prejudice.

Demographic variables.

The attribute variables for this study included age, gender, ethnic origin, educational level, socio-economic status as reflected by type of health insurance, current employment or student status, reported genotype of SCD, relationship status, age of SCD diagnosis, last sickle cell pain crisis, number of reported vaso-occlusive crises (VOCs) in the past 12 months that were treated in emergency department only as well as were treated as a patient in the hospital, and reported diagnosis of other SCD complications.

Methods and Procedures

Setting

The setting for this study included a global population of adult persons diagnosed with SCD or sickle cell anemia. In America, it is estimated that 100,000 Americans are impacted by this chronic illness. In the United States, one out of every 365 African Americans and one out of every 16,300 Hispanic-Americans are diagnosed with SCD (CDC, 2016). Miami-Dade County was presumed to be the starting place to recruit participants, however due to the COVID-19 pandemic the use of social media became the setting. In Miami-Dade county, there was a total population of 2,693,117 persons where 18.7% of the population were African-American and 66.8% were Hispanic-American. These statistics are notable because African Americans and Hispanics have a higher incidence of SCD than Caucasions. There were approximately 4,294 sickle cell patients enrolled in the Florida Medicaid program (Reuters Health, 2009). However, it is unknown how

many lived in Miami-Dade county. According to the United States Census Bureau (2014), the population within Miami-Dade county had a median household income of $43,009.00 with 20.4% classified at the poverty level. The educational breakdown in Miami-Dade county was 86% with a high school diploma or higher; and, 29% with a bachelor's degree or higher (United States Census Bureau, 2014). Among the population that was under age 65 years in Miami-Dade county, 37% were unemployed, and 11% were without health insurance. Eighty-six percent of the population in Miami-Dade county were 18 years old and older (United States Census Bureau, 2014).

Sample

A convenience sample of adults with SCD was recruited. The minimum number of participants required to participate was determined based on a moderate effect size (r=.15), α=0.05, and power=0.80, and with a maximum number of predictor variables= 5. A total of 91 participants were determined to be an appropriate sample size for this study.

Study Population

The population for this study was anyone self-reporting a medical diagnosis of sickle cell disease or sickle cell anemia, aged 18 years old and older, who could read, write and speak English. Additionally, persons of any race, educational level, employment status or socio-economic status, who reported a diagnosis of sickle cell anemia or sickle cell disease were eligible to participate in the study.

Inclusion Criteria.

Only persons with a self-reported diagnosis of sickle cell disease of any genotype were asked to participate. Participants were able to comprehend and speak English. It was not necessary for participants to read because all consents and interviews were read by the PI. All interviews were conducted via telephone due to COVID-19 CDC guidelines, which emphasized social distancing thereby limiting face-to-face interviews. Interpreters were not used for any interview.

Exclusion Criteria.

Persons with sickle cell trait were not eligible to participate. Participants who are diagnosed with sickle cell trait do not experience sickle cell symptoms unless in extreme circumstances of oxygen deprivation and dehydration. Hence, they are not likely to experience sequelae from SCD. Also, any individual who was diagnosed with cognitive and developmental problems that prevented them from providing informed consent or that prevented them from accurately understanding the study questionnaire was excluded from the study. Lastly, persons experiencing an active incapacitating SCD pain episode or “crises” was deferred from data collection until they were able to comfortably participate in telephone interviews. Participants experiencing active incapacitating sickle cell pain could find the pain so disruptive it could interfere with the quality and accuracy of their responses.

Recruitment of Participants

After approval from Florida International University’s Institutional Review Board (IRB), the recruitment process was initiated. Flyers that included a brief

description of the study and the contact telephone number of the researcher were provided to interested participants.

Due to the pandemic of COVID-19, flyers were not placed at local SCD support group meetings, doctor's offices, and clinics where adult SCD patients are treated because of CDC guidelines to socially distance. The PI contacted administrators of several social media groups for permission to upload flyers. Flyers were emailed to social media administrators who uploaded them in their social media timelines. The social media support groups were *Advancing Sickle Cell Advocacy Project, Inc*., *Sickle Cell Anemia Disease,* and *Sickle Cell Consortium*.

Also, due to the pandemic, there were many virtual support group meetings and SCD conferences attended by the PI. The conferences attended were the *12th Annual Sickle Cell Disease Summit*, the *SCDAA 48th Annual National Convention*, and the *7th Annual Sickle Cell Patient and Family Symposium*. During virtual support group meetings and SCD conferences, the PI asked for participation either via zoom or from the email contact lists of participants provided by the conferences. Additionally, the "snowball recruitment method" was employed by asking participants who were interviewed or acquaintances of the PI to inform persons with SCD about the study. Participants or acquaintances of the PI who identified participants willing to complete the survey were provided a flyer, by either email or text, or the PI's telephone number for the new participant. When new

participants were identified, telephone interviews were scheduled at the convenience of the participants.

Data Collection

For participants who agreed to participate and provided contact information, the PI scheduled an appointment at the participants' convenience. At the scheduled time, the PI read the consent, give a description of the study, then asked every survey questions, 99 in total. Some responses were written on a printed copy and some were entered directly into Qualtrics. Time was allotted for participants to completely express their feelings. Upon completion of the survey, the PI thanked the participant for their time and inputted the written responses in Qualtrics. If depression was not identified, the printed copy with the participant's name, phone number, and email address was placed in a secured location only accessible to the PI. When participants expressed a desire to complete the survey on-line, their name, contact phone number, and email address was collected, the link to Qualtrics was provided via email. The consent was a part of the survey in Qualtrics, participants had to provide consent to enter the survey. The PI has the only access to survey in Qualtrics, the only identifying information in Qualtrics was their email address. The PI has the names, email addresses, and phone numbers of participants which are placed in a secured place only accessible to the PI. The PI analyzed BDI depression immediately after telephone interviews or in Qualtrics. Participants who were determined to be mild mood disturbance, borderline clinical depression, moderate depression, severe depression, and extreme depression based on Beck's Depression Inventory was referred to local mental health

providers, only if participant is not receiving treatment. Participants via Qualtrics determined to be mild mood disturbance, borderline clinical depression, moderate depression, severe depression, and extreme depression were notified by PI via email or telephone and referred to local mental health providers for evaluations, only if participant is not receiving treatment. Most participants who were determined to exhibit depression were aware of their conditions and were receiving mental health services.

Instrumentation

This research required each participant to complete five questionnaires that measure the key constructs of the study and a demographic questionnaire. The five questionnaires and demographic questions were organized and inputted into Qualtrics entitled "Chronic Disease Outcome Triad". The questionnaires that were included are the following: Demographic Questionnaire, Brief Pain Inventory (BPI), SCD Symptomatology Questionnaire, Beck's Depression Inventory (BDI), Chronic Disease Attitude Scale, and the Sickle Cell Self-efficacy Scale (SCSES).

Demographic Questionnaire:

This questionnaire was designed to obtain background data on the demographic characteristics of each participant such as age, gender, race, educational level, type of insurance, genotype, marital/relationship status, employment status and/or school status, age at SCD diagnosis, last pain crisis, number of hospital and emergency department visits for pain crises in past 12 months, and other SCD complications.

Measurement of Triadic Sequelae Domains

Brief Pain Inventory (BPI)

This instrument was used to measure pain as an indicator of the physiological triadic domain. This instrument has been used extensively in cancer patients and is increasingly used as an outcome measure in patients with other chronic diseases (Gjeilo et al., 2007). The BPI is a multidimensional measure that consists of questions related to pain severity and questions related to pain interference on function. In this study, participants rated their pain severity and interference over the past 30 days. Since sickle cell pain is often frequent, intermittent and unpredictable, 30 days allowed for a more narrowed perspective of their experience of pain. The instrument uses a scale of 0=”no pain” to 10=”pain” as bad as it can be” and allows for the assessment of pain at its worst, least, and average. The BPI reports two main scores: a pain severity score and a pain interference score. The pain severity score is calculated from four items and scores range from 0 to 40. The pain interference score is calculated from seven items and scores range from 0 to 70.

In a study that investigated the prevalence of insomnia symptoms and which identified biophysical predictors in community-dwelling adults (18 years and older) diagnosed with SCD, the BPI was used to measure acute pain and the degree to which pain interfered with daily functioning and feelings (Moscou-Jackson et al., 2016). The Cronbach’s alpha for the BPI in this study was 0.87. In a study that investigated acute pain in patients receiving cardiac surgery, pain assessments before surgery and six-months after surgery resulted in an alpha coefficient of 0.84

for severity and 0.91 for interference before surgery; and, 0.89 for severity and 0.94 for interference at a six-month follow up item-to-total correlations indicated that each item contributed to the construct it is intended to measure and is reliable (Gjeilo et al., 2007). In the cardiac surgery study (Gjeilo et al., 2007), the criterion validity was supported by correlations, between the BPI pain severity index and the SF-36 BP scale, which was -0.47; and, correlations between the BPI pain interference index and the SF-36 BP scale was -0.53.

SCD Symptomatology Questionnaire

This questionnaire (Edwards, Telfair, & Lenoci, 2001) has two subscales: the SCD Physical Symptoms Subscale and the SCD Psychological Symptoms Subscale.

SCD Physical Symptoms Subscale

This questionnaire was used to measure physical SCD symptoms in addition to pain to reflect the overall physical state of the individual as another indicator of the physical triadic domain. This measurement required patients to rate the frequency of 11 physical symptoms. The 11 physical symptoms are weakness, yellowing of skin or eyes, vomiting, nausea, pain, heart problems, gall stones, eye trouble, kidney problems, swelling of hands or feet, and shortness of breath (Edwards, Telfair, & Lenoci, 2001). For the 11 physical symptoms the response categories include 1=never or rarely (zero or one time), 2=not very often (two or three times), 3=often (four or five times), or 4= very often (six or more times) (Edwards, Telfair, & Lenoci, 2001). This measurement tool was used in a study

investigating self-efficacy as a predictor of adult adjustment which indicated a Cronbach's alpha of 0.81 for physical symptoms, thereby signifying good Internal consistency (Edwards, Telfair, & Lenoci, 2001).

SCD Psychological Symptoms Subscale

This instrument was used to measure multiple psychological symptoms in order to reflect the overall mental/emotional state of the individual as an indicator of mental/emotional triadic domain. This measurement required the patients to rate the frequency of eight psychological symptoms over the preceding six-months. The eight psychological symptoms are feeling sad, feeling tense or nervous, feeling short-tempered, feeling worried or concerned, problems coping, problems sleeping, problems eating, and problems paying attention (Edwards, Telfair, & Lenoci, 2001). For the eight psychological symptoms, the response categories for items comprising this measure is the same as physical symptoms (1-4) (Edwards, Telfair, & Lenoci, 2001). For both categories all responses were added up to determine the overall psychological indexes and greater scores indicated higher levels of reported psychological symptomatologies (Edwards, Telfair, & Lenoci, 2001). This measurement was used in a study investigating self-efficacy as a predictor of adult adjustment which resulted in a Cronbach's alpha of 0.84 for psychological symptoms, signifying good internal consistency (Edwards, Telfair, & Lenoci, 2001).

Beck's Depression Inventory (BDI)

This instrument was used to measure depression as an indicator of the mental/emotional triadic domain. Depression assessment is important in the study of SCD to indicate the emotional well-being of the individual. This questionnaire includes 21-items corresponding to symptoms of depression (Beck et al., 1961). Each item is totaled to give a single score for the BDI (Beck et al., 1961). There is a four-point scale for each item which is scored ranging from 0-3 (Beck et al.,1961). Total scores can range from 0-63. Scores for level of depression are: 0-10 the ups and downs are considered normal, 11-16 mild mood disturbance, 17-20 boderline clinical depression, 21-30 moderate depression, 31-40 severe depression, and over 40 extreme depression (Beck et al., 1961). The BDI has over 35 years of evidence for its reliability and validity (Gibson et al., 2013; Edwards et al. 2009; Jenerette, Funk, & Murdaugh, 2005). It has been shown to be highly reliable and valid regardless of the population. Its average coefficient alpha is .80 or above and its construct validity has been supported consistently differentiating depressed from non-depressed patients (Gibson et al., 2013; Edwards et al., 2009; Jenerette, Funk, & Murdaugh, 2005).

Behavioral Health Outcomes

The behavioral outcomes that was studied within the context of this study are sickle cell fatalism, perceived SCD prejudice, and sickle cell self-efficacy. These constructs were measured using the Chronic Disease Attitude Scale to measure sickle cell fatalism and perceived SCD prejudice, and the Sickle Cell Self-efficacy Scale to measure sickle cell self-efficacy.

Chronic Disease Attitude Scale

The Chronic Disease Attitude Scale is comprised of two subscales: (1) the Chronic Disease Fatalism Subscale, and (2) the Perceived Chronic Disease Prejudice Subscale (Strickland, 2013). The conceptual base of the Chronic Disease Attitude Scale was derived from the results of a qualitative study that explored quality of life of persons with SCD (Strickland et al., 2001), which showed the dominant role of two negative chronic disease attitudes, i.e., fatalism and perceived prejudice, had on the wellbeing and quality of life of persons with SCD. In addition, the literature was searched for other descriptions of these constructs in relation to other chronic diseases. The questionnaire was then designed to measure the two predominantly identified chronic disease attitudes, i.e. chronic disease fatalism and perceived chronic disease prejudice. *Chronic disease fatalism* is the belief that one has no control over the symptoms associated with the chronic disease and its related complications, and that those with the chronic disease will have an early death (Strickland, 2013). *Perceived chronic disease prejudice* is the belief that others view persons with the chronic disease negatively or in a biased stereotypical way because they have the disease (Strickland, 2013). The questionnaire was designed for completion by persons diagnosed with a specific chronic disease, or by others who are not diagnosed with the disease, but for whom one may want to measure attitudes about a specified chronic disease (Strickland, 2013).

Most of the items for the Chronic Disease Fatalism Subscale were derived from the literature, i.e., examples from the health-related literature on chronic

diseases were used to generate items. Some of the items on the Cancer Fatalism Scale (Powe,1996), which has very good evidence for reliability and validity, were adapted to measure fatalism on the Chronic Disease Attitude Scale (Strickland, 2013). Items were written at a 5th grade reading level (Strickland, 2013). The format of the Chronic Disease Attitude Scale was developed with a similar format as the Cancer Fatalism Scale (Strickland, 2013). Items are declarative statements to which respondents are asked to rate their level of agreement or disagreement along a Likert scale which ranges from 0 to 10 (Strickland, 2013). Directions instruct the respondent to "circle the number on the scale from 0 (agree) to 10 (disagree) that best shows how you feel" (Strickland, 2013). The name of the specific chronic disease that is the focus of the study can be specified in the questionnaire if this enhances clarity (Strickland, 2013). The Chronic Disease Attitude Scale is scored by summing item scores for each of the subscales. Therefore, low scores indicate lower amounts of fatalism and perceived prejudice and higher scores represent higher amounts of the constructs related to the specified disease (Strickland, 2013).

The questionnaire items have *a priori* content validity because they were developed from the questionnaire's explicated conceptual base, responses from a qualitative study (Strickland et al., 2001) that described the nature of the measured constructs, and the published literature (Waltz, Strickland, & Lenz, 2017). Since some items for the Chronic Disease Attitude Scale were adapted from a previously validated measure of fatalism (Powe,1996), the questionnaire has additional support for its validity (Waltz, Strickland, & Lenz, 2017). *Posteriori* content validity

was also assessed based on ratings of two content experts. A content validity index of 1.0 was derived for the Chronic Disease Attitude Scale and its subscales. This measurement tool is a 20-item instrument with response choices that range from zero to ten, with 0="disagree", 5="do not agree or disagree", and 10="totally agree." This instrument has a content validity score of 1.0 based on the ratings of two content experts. This study was the first to use the tool with a SCD population.

Sickle Cell Self-Efficacy Scale (SCSES)

This instrument was developed to assess participants' perception of their ability to function on a day-to-day basis and manage SCD's symptomatology (Edwards et al., 2000). This instrument measures self-efficacy using 9-items with response choices ranging from "not at all sure", "not sure," "neither", "sure," or "very sure" (Edwards et al., 2000). Scores are obtained by adding item scores where higher scores indicate greater self-efficacy. The SCSES demonstrated good internal consistency (0.89), discriminant validity, convergent and predictive validity (Edwards et al., 2000).

Data Analysis

All data were analyzed using the IBM SPSS version 27.0 (2020). After data was collected, data was placed in Qualtrics . When data collection was completed, it was transported into SPSS. The variables representing the triadic sequelae, behavioral health outcomes, disease-related characteristics, and characteristics of participants were re-named and recoded as necessary before analyses were begun. The approach to data analyses began with basic descriptive analysis, which included the calculation of means, standard deviations, and ranges for all

interval and ratio level data. For interval and ordinal level data, Pearson's correlation coefficients, Point-biserial, correlations Canonical correlations, and Spearman's rank-order were calculated between all variables to assess their intercorrelations. Data that are categorical and ordinal were analyzed using frequencies, modes, and ranges. Preliminary analyses were done to determine if the data conform with the assumptions that serve as the basis for statistical analyses. Additionally, Stepwise hierarchal multiple regressions were conduted to determine the predictive abilities of multiple variables.

The data analysis procedures were presented based on the study hypotheses and research questions. Each study hypothesis or question was presented, followed by the statistical approach to analysis.

Specific Aim 1

To assess the relationship between the number and severity of SCD sequelae in each of the triadic domains with each other and as predictors of behavioral health outcomes.

Hypotheses:

1. There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores, and a negative relationship of these two domains with the sociocultural domain scores when domain variables are assessed both individually and categorically.

Individual variables within each domain was correlated with each other based on the levels as specified. Number and severity of variables within each

domain was analyzed using Pearson's product moment correlations. Point-biserial correlations were used to determine the relationships between dichotomous variable sociocultural domain and continuous variables physiological domain and psychological domain. This required several separate analyses.

2. There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores with negative behavioral outcome scores, and a negative relationship with positive behavioral outcome scores; and, there will be a negative relationship of the higher sociocultural domain score with negative behavioral health outcome scores and a positive relationship with positive behavioral outcome scores.

A Pearson's product moment correlation was used to analyze the relationship between predictor variables (physiological sequelae and psychological sequelae) and criterion variables (sickle cell fatalism scores (Fatalism), perceived SCD prejudice scores (Perceived Prejudice), and SCD self-efficacy scores (SCSES)). There were six separate analyses with each predictor variables and each criterion variables. A Point-biserial correlation was used to analyze the relationship between predictor variable (sociocultural variables; employment and school) and criterion variables (Fatalism, Perceived Prejudice, and SCSES). There were three separate analyses.

3. There will be a positive relationship between negative behavioral outcome scores (Fatalism, Perceived Prejudice) with each other, and an inverse

relationship with positive behavioral health outcome scores (SCSES) in adults with SCD.

Pearson's cproduct moment and Spearman's rank order correlation were used to analyze this hypothesis. There were three separate analysis.

4. The number and severity of triadic domain scores will predict negative and positive behavioral health outcome scores.

Multiple linear regressions were used to analyze this hypothesis. Each triadic sequelae domain score was analyzed with each behavioral health outcome scores. This required nine separate analyses.

Specific Aim 2:

To examine the relationship of demographic and disease-related characteristics of adults with SCD as predictors of triadic sequelae and behavioral health outcomes of SCD.

Hypotheses:

1. The more frequently adults with SCD report painful episodes over the past year, and the presence of diagnosed SCD complications, and hospitalizations/emergency department visits over the past year; the higher will be the number and severity of physiological and psychological domain scores, and the lower will be the sociocultural domain scores.

This hypothesis was tested with the Pearson's product moment correlation analysis. Each predictor variables (SCD complications, PainER, and PainHosp)

was analyzed separately with each criterion variables (physiological and psychological sequelae). A point-biserial correlation analyzed each predictor variables (SCD complications, PainER, and PainHosp) and criterion variable (sociocultural variables).

2. Older age at diagnosis, older current age, and female gender will be positively associated with positive behavioral health outcomes (SCD self-efficacy) and negatively associated with negative behavioral health outcomes (SCD fatalism and perceived sickle cell prejudice).

This hypothesis was tested using Pearson's correlations. The predictor variables Diagnosed Age and Current Age were analyzed with each criterion variables SCSES, Fatalism, and Perceied Prejudice. The predictor variable female gender was tested using point-biseral correlations with each criterion variable.

Research Questions:

1. To what degree do the number and severity of physiological and psychological domain scores, and the sociocultural domain score predict reported painful episodes, diagnosed complications, and hospitalizations/emergency department visits over the past year?

This question used multiple linear regressions for its analysis. There were multiple analyses performed using physiological sequelae, psychological sequelae, and sociocultural variables as the predictor variables

and reported SCD Complications, PainER, and PainHosp as the criterion variables. Each predictor variable were analyzed with each criterion variable.

2. To what degree do older age at diagnosis, older current age, and female gender predict behavioral health outcomes, (i.e., SCSES, Fatalism, and Preceived Prejudice)?

This hypothesis used multiple linear regressions to determine if the predictor variables of Diagnosed Age, Current Age, and Female gender can predict criterion variables SCSES, Fatalism, and Perceived Prejudice.

Human Subjects Consideration

After obtaining approval from Florida International University's (FIU) IRB, the PI promoted responsible conduct of research by practicing high standards of ethics and accountability in planning, implementing, and informational distribution. Additionally, the PI adhered to behavioral standards according to FIU by providing a safe data collection environment that included privacy and support when taking the survey.

Due to the COVID-19 pandemic, there were no face-to-face interviews. Therefore, there were no written questionnaires from the participants, interviews were conducted via telephone with the PI. Each questionnaire was labelled with their name and kept private by the PI. Therefore, participant's names were protected and not entered into data collection files. All completed questionnaires are kept in a locked box and file cabinet located within a locked office. Completed questionnaires were entered into Qualtrics, only email addresses were included.

Inclusion criteria and exclusion criteria were strictly adhered to during the recruitment and data collecting processes. Participants were advised that their participation requires completion of questionnaire one-time only and will approximately take 45 minutes to 60 minutes to complete.

Sources of materials.

The materials for this study were questionnaires compiled together into one survey entitled “Chronic Disease Outcomes Triad” which were completed by either a telephone interview with the researcher or via online in Qualtrics.

Potential risks.

Potential for injury from participation in this study was minimal. Completion of telephone interviews or on-line surveys were free of risks for physical harm to participants or breach of confidentiality because the data will only be assessable to researcher. Data was electronically transported from Qualtrics to SPSS by researcher only. There were no questions that caused emotional upset known to the researcher.

Adequacy of Protection against Risks

Informed Consent.

When telephone interviews were conducted, the PI read the consent to participants, and upon agreeing with the consent, the survey began. Due to the pandemic of COVID-19, there were no face-to-face support groups meeting, well-clinic visits, or personal meeting places. Support group meetings and conferences were virtual and the researcher informed potential subjects about the study, offered

them the opportunity to participate, answered any questions about the study, and obtained their contact information. The PI's email address and contact number were provided to participants answering the survey in Qualtrics. It was emphasized to the participants that their participation was completely voluntary and that they had the right to withdraw at any time without any penalty or threats to access to their healthcare. Participation was completely confidential and voluntary since data was deidentified and kept in locked files. Also, on-line surveys had email addresses only and is kept in a secured file only accessable to the PI. Participants gave informed consent to participate either via telephone or on-line.

Adherence and retention procedures.

Individuals who agreed to participate in the study was required to answer survey with PI in one telephone interview or in Qualtrics. All participants who agreed to telephone interviews were scheduled at their convenience. The PI used positive communication strategies that are socially and culturally supportive.

Procedure to minimize risks.

Risks for injury were minimal as previously mentioned. However, a potential risk in this study could be breaks in confidentiality. All hard copies that contained confidential information such as informed consents and questionnaires are kept in a locked box with access only to the PI in a separate file cabinet.

Potential benefits of the proposed research to the subjects and others.

Individuals in this study will gain no direct benefit from this study beyond self-sacrificing rewards for participating in the development of science in this topic area.

Importance of knowledge to be gained.

This research will significantly impact the healthcare communities, and persons with sickle cell disease by reporting the relationship of physiological, psychological, and sociocultural sequelae, chronic disease attitudes, and self-efficacy. These relationships may profoundly impact the self-management and worldview of individuals living with this condition. Additionally, the reporting of the relationships may educate the health care communities and the communities in general on how to properly treat and respect adults with SCD. An accurate investigation of this magnitude is an important step initiating further research on the development of chronic disease attitudes and self-efficacy and how they impact patients with SCD and other PCMIs.

Inclusion of women, minorities, and children.

Inclusion of women. Women were included in the study. It is anticipated that 50% of the participants will be women. The reliability and validity of the Chronic Disease Outcome Triad Study is not based on gender because both genders have SCD.

Inclusion of minorities. The sample will include primarily individuals with African and Latino backgrounds. Approximately 99% of persons diagnosed with SCD have an African or Latino heritage.

Inclusion of children. Children are not included in this study. Participants must be 18 years and older.

Data sharing plan.

At the end of this project, the PI will prepare a data set, stripped of identifiers, that could be made available to other researchers.

Assumptions and Limitations of the Study

Within the context of the study, data was collected from volunteers with SCD who served as respondents to survey questionnaires. Major methodological assumptions of the study are that persons who volunteer to participate in the study will be generally representative of those who have SCD and that their responses to all questionnaires regarding their health and related experiences will be honest and truthful. The researcher encouraged respondents to answer truthfully by ensuring participants that their responses will be confidential and their participation will remain anonymous. Only survey questionnaires that have prior evidence of their reliability and validity were used in the study to ensure that the data collected was as reliable and valid as possible. However, this study is the first one conducted using the Chronic Disease Attitude Scale with persons with SCD. Although the instrument was designed based on a well-developed cancer questionnaire and the literature which gave sound content validity, its limited use in this population

requires detailed assessment of its reliability and validity in SCD patients. The known reliability and validity of the Chronic Disease Attitude Scale is based on nursing students and health-administration students. The study also tests the researcher's newly developed Chronic Disease Outcomes Triad conceptual framework on an actual population with a chronic disease.

Also, questionnaires was the primary method of data collection and therefore findings were based primarily on patients' perceptions. It was assumed that participants answered all questionnaire items honestly and without bias. The degree to which this assumption is correct is unknown. Although steps were taken to encourage truthfulness, reliability, and validity of the data collected, use of volunteers as respondents and the collection of data using self-report questionnaires are limitations of the study. Generalizations of findings of the study were done with these limitations in mind, particularly since those who volunteer to participate in research may be a somewhat different population subset and have different perspectives than those with SCD who chose not to participate.

Summary

This study was the first step in reporting consequences of the inter-relational and interactional connection of physiological, psychological, and sociocultural sequelae, known as the triadic sequelae for predicting positive and negative behavioral outcomes. This study will be instrumental in providing information to health care providers and the community that will inform them of the impact of chronic diseases and painful chronic medical illnesses.

This chapter presented the research design and methodology for this study. A comprehensive description of the research design which included predictor, criterion, and demographic variables were reported. Information on methods and procedures which included the setting, study population, sample with sample size calculation, inclusion and exclusion criteria, recruitment techniques and data collection were described. A thorough description of all study instruments and available reliability and validity data was given. Data analysis techniques for each hypotheses and research questions were provided.

IV. Research Results

Overview

This study examined the inter-relational and interactional relationships that patients may experience when they have a chronic illness, i.e. physiological sequelae, psychological sequelae, and an important sociocultural variable (employment/school involvement) that are relevant aspects of the triadic sequelae of sickle cell disease (SCD) that are likely to be associated with SCD outcomes. This study investigated the relationship of and predictiveness values of key triadic sequelae on behavioral health outcomes. Also, the study investigated the predictive values of other characteristics of adults with SCD, such as age at diagnosis, pathophysiological aspects of SCD (i.e. genotype), and debilitating effects of the disease such as diagnosed SCD complications (i.e. stroke, avascular necrosis (AVN), and eye problems) on behavioral health outcomes. Within the context of this study, the triadic sequelae studied for the physiological domain included, the Brief Pain Inventory-short form (BPI) scores (Cleeland, 1989) and SCD symptomatology-physical which were separated to include the presence and frequency of the 11 symptoms (Edwards, et al., 2001) scores. The sample means (SD) for Physical were 22.93(4.75) and frequency (F-Physical) was 5.68(1.93). The study's sample means and standard deviation of the variables from BPI, included Pain Severity 17.19 (6.08) and Pain Interference 34.91 (15.82) (Cleeland, 1989). For the psychological domain, depression was measured using the instrument Beck's Depression Inventory (BDI) (Beck, et al., 1961) and SCD symptomatology-psychological scores (Edwards, et al., 2001) which was also

separated to include the presence and frequency of the 8 psychological symptoms were investigated. The sample's mean (SD) for BDI was 10.58(8.60), Psychological was 17.37(6.20), and F-Psychological was 4.79(2.36). For the sociocultural variables, employment status and/or school attendance were obtained using the Demographic Tool. The responses of employment and school attendance were combined to create one score. There were 39(42%) who were not working or attending school and 53(58%) who were working and/or attending school. The behavioral health outcomes researched in this study were both negative and positive. The means (SD) of negative behavioral health outcomes studied were Sickle Cell Fatalism (Fatalism) 37.30(14.02) and Perceived Sickle Cell Prejudice (Perceived Prejudice) 21.84 (10.69). The mean (SD) of positive behavioral health outcome variable Sickle Cell Self Efficacy (SCSES) was 33.93(5.60). The testing of the Chronic Disease Outcomes Triad (CDOT) Model, which was developed within the context of this study and focuses on the triadic sequelae and their predictive nature on behavioral health outcomes in SCD was of particular interest. The purpose of this chapter is to report the results of the data analyses related to the research hypotheses and questions. Results are presented in several sections. The first section begins with a comprehensive view of the demographic characteristics of the 93 adult participants in the study who were diagnosed with sickle cell disease who were 18 years and older at the time of the study. Study participants included persons self-reporting any genotype of SCD, any ethnicity, and any gender.

Recruitment, Screening, and Data Collection

The study received IRB approval from the Florida International University Institutional Review Board in April 2020, which was at the beginning of the COVID-19 pandemic. Therefore, due to the pandemic, the PI had to recruit participants virtually and via social media, rather than face-to-face due to high risks of transmitting COVID-19 via physical contact during the pandemic. The PI contacted administrators of several social media groups and received permission to upload recruitment flyers. Flyers were emailed to social media administrators who were willing to assist the PI by uploading the flyers in their social media timelines. The social media support groups that uploaded flyers were *Sickle Cell Anemia Disease*, the *Sickle Cell Consortium*, and *Advancing Sickle Cell Advocacy Project, Inc.* Social media support group membership ranged between 200 to 25,000 members per group, however, the response rate was very low (n= 9).

In addition, due to the pandemic, there were several virtual support group meetings and SCD conferences attended by the PI. The conferences the PI attended were the *12th Annual Sickle Cell Disease Summit, SCDAA 48th Annual National Convention*, and the *7th Annual Sickle Cell Patient and Family Symposium*. The support group that the PI attended monthly was *Advancing Sickle Cell Advocacy Project, Inc*. that convened bimonthly. During virtual support group meetings and SCD conferences, the PI asked for participation either via zoom or from the email contact lists of participants provided by the conferences. The response rate using this method was more successful when the PI was able to communicate directly with the participants (n = 60). Additionally, the PI employed

the “snowball recruitment method” by asking participants who were interviewed or acquaintances of the PI to inform persons with SCD about the study. For participants or acquaintances of the PI who identified respondents willing to complete the survey, a flyer or the PI’s telephone number was provided, by either email or text, to the prospective participant. The responses using this method was successful when the PI directly communicated with the participants (n = 24). Additionally, participants who received the Qualtrics link without verbal communication with the PI, had a very low response rate where participants either partially completed or failed to complete the survey (n = 29). There were 122 survey responses with approximately a 76% completion rate (n = 93). All telephone interviews were scheduled at the convenience of the participants and conducted by the PI. The PI read the consent form to each participant and the interviews were conducted via telephone. Interview times ranged between 40 minutes to 150 minutes. After the interviews were completed, the participants were thanked for their time.

Data Analysis Procedures

Data for the 93 study participants were analyzed using the IBM SPSS 27.0 version software (2020). Each statistical hypothesis was tested based on a 0.05 significance level. General guidelines for the interpretation of correlations were as follows: 0.1 to 0.3 = small effect; 0.3 to 0.5 = moderate effect; and 0.5 and higher equal a large effect. Chapter three presented the reliability statistics that were calculated in the sample for the specific instruments. Scatterplots, casewise diagnostics, and studentized deleted residuals (SDR) were visualized and

analyzed to explore the relationships between variables and identify outliers. Shapiro-Wilk's test, Histograms with superimposed normal curve, P-P Plots, and studentized residuals (SRE) were used to determine normality. Levene's test of homogeneity, correlation coefficients, and Tolerance/VIF were used to assess for multicollinearity. Leverage points and Cook's Distance were used to assess for unusual points. Means, standard deviations, frequencies, Pearson's product-moment correlations, Spearman's rank-order correlations, Canonical correlations, Hierarchal regressions, and general linear models were calculated according to the nature of the data, specific aims, and hypotheses. Missing data were not included in computations of the variable scores. Separate analyses were required for the examination of each hypothesis. Analyses for research questions were specified in the previous chapter and will be revisited in this chapter as results are presented.

Description of the Sample

The study sample was recruited virtually or via email which afforded a global population with 122 potential participants responding to recruitment efforts. There were no face-to-face data collection meetings due to the COVID-19 pandemic therefore interviews were conducted via telephone. The participants were males and females, between the ages of 18 to 74 years old, self-reporting any genotype of SCD, and able to understand and speak English. Most participants were living in the United States at the time of data collection, however, there were 22 participants from different countries who participated in this study. (See Tables 1-5).

Table 1

Country of Origin of Participants, including missing data (N=122)

Country	Frequency	Valid Percent
Bahrain	5	4.1
France	2	1.6
Ghana	2	1.6
Ireland	1	.8
Kingdom of Saudi Arabia	2	1.6
Malaysia	1	.8
Nigeria	3	2.4
Oman	1	.8
Uganda	1	.8
United Kingdom-London	4	3.3
United States	76	60.8
Missing	24	21.4
Total	122	100

Table 2

Gender of Participants (n=93)

Gender	n	%
Male	29	31.2
Female	64	68.8
Total	93	100

Table 3

Participants' Ages in Categories (n=93)

Age	n	%
18-24	5	5.4
25-34	27	29
35-44	33	35.5
45-54	14	15
55-64	9	9.7
65 and over	5	5.4
Total	93	100

Table 4

Ethnicity (n=93)

Ethnicity	n	**%**
Black, African American, or African	81	87
Asian/Pacific Islander	2	2.2
Hispanic or Latino	4	4.3
Other	6	6.5
Total	93	100

Table 5

Self-reported Sickle Cell Genotype (n=93)

Genotype	n	%
HbSS	64	68.9
HbS/b-O thalassemia	3	3.2
HbS/b+ thalassemia	8	8.6
HbSC	18	19.4
Total	93	100

Characteristics of the Study participants

A demographic questionnaire was created to collect data on the participants' gender, age, ethnic origin, educational level, insurance coverage, genotype of SCD, relationship status, employment status, school status, age at diagnosis of SCD, and last sickle cell pain crises. Additionally, more questions collected data on the number of times a participant was treated in the emergency room only for sickle cell pain crises in the past 12 months, and the number of times a participant was admitted in the hospital for sickle cell pain crises in the past 12 months. Lastly, the participants were asked if he or she was diagnosed with other SCD complications, such as stroke, AVN, and eye problems. Tables 6 and 7 report these data. Only 17 (18.3%) had no diagnosed SCD complications. Therefore, 81.7% of the sample had at least one complication. The category with the highest number of participants was those with two SCD complications, with 17.3% of participants experiencing two complications. Note that most participants with complications had between two to five complications. Eighteen participants or 19.4% had six or more complications.

Table 6

Number of SCD Complications

SCD Complications	Number of Complications	*f* (%) of participants with Number of Complications
	0	17(18.3)
	1	12(12.9)
	2	16(17.2)
	3	10(10.8)
	4	11(11.8)
	5	12(12.9)
	6	7(7.5)
	7	4(4.3)
	8	3(3.2)
	9	0(0)
	10	1(1.1)

Table 7 notes that the most frequently reported SCD complications were: avascular necrosis, painful and swollen joints, eye problems, lung complications, gallstones, stroke, enlarged spleen, heart problems, and kidney disease. Most of these in most circumstances would be considered serious complications.

Table 7

Types of SCD Complications and number of participants with complications

SCD Complications	*f*	%
Stroke	22	24
Enlarged Liver	6	7
Kidney Disease	12	13
Painful and swollen joints	35	38
Leg Sores	8	9
Heart problems	15	16
Lung Complication	29	32
Pulmonary Hypertension	8	10
Avascular necrosis (AVN)	36	44
Restless Legs Syndrome	7	9
Priapism	9	11
Mental illness	11	17
Eye Problems	30	46
Enlarged spleen	19	33
Ear Problems	8	14
Gallstones	27	47

There were 38 (41%) adults with SCD who reported some college education but did not receive a bachelor's degree. This category represented the respondents who either had an associate degree or attended college but did not graduate. Socioeconomic status was determined in this study based on whether

participants had insurance. Thirty-two percent of the participants had Medicare and 11% had no insurance. There were more adults with SCD who reported being "single", "divorced", "separated", or "widowed" (58%) than "married" or "in a relationship" (42%). Additionally, 52% of this sample were working and only 13% were attending school at the time of the study. Of the sample, 84 (90.3%), were diagnosed with SCD from birth to less than 7 years old. Several of the participants, i.e. 27 (29%) reported that their last pain crisis was either less than one week from the interview or between one to three months prior to the interview. Only 11 (12%), were treated for sickle cell pain in the emergency room five or more times in the past 12 months. Additionally, only 12 (13%), were admitted to the hospital for sickle cell pain five or more times in the past 12 months.

Table 8

ER Visits and Hospital Admissions for Sickle Cell Pain Crisis in 12 months ƒ(%) (n = 92)

Number of Visits	0 -1	2-4	5 or more
ER	64(69.6)	17(18.5)	11(12)
Hospital	61 (66.3)	19(20.7)	12(13)

Results of Data Analyses

Results of data analyses for each of the study aims and their related hypotheses and research questions are presented in this section. Initially, data were checked for violations of assumptions prior to performing all statistical tests. Variables were analyzed using Pearson's product-moment, Spearman's rank order, and Canonical correlations to examine the relationships between

continuous and/or categorical variables. Point-biserial correlations analyzed dichotomous variables. Correlations were examined using a two-tailed approach unless directional relationships were hypothesized and results were in the hypothesized direction. In such cases, results were interpreted using the one-tailed approach with appropriate adjustments in reported directional probabilities. Since most of the independent variables were significantly correlated with each other, hierarchal stepwise multiple linear regressions analyses were conducted to address hypotheses that investigated the predictive values between multiple independent variables and specified dependent variables. The general linear model analyzed hypotheses that investigated the predictive values and 95% confidence intervals were used for relationships between independent and dependent variables. Findings for each hypothesis are organized and presented according to their associated major aims. Each aim's, related hypotheses, and research questions are restated prior to presenting their associated results to enhance clarity.

Research Aims and Questions

Aim 1.

To assess the relationship between the number and severity of SCD sequelae in each of the triadic domains with each other and as predictors of behavioral health outcomes.

Hypothesis 1. There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores, and a

negative relationship of these two domains with the sociocultural domain score when domain variables are assessed both individually and categorically.

Physiological Sequelae. A Pearson's product-moment correlation was run to assess the relationship between the variables used for the physiological domain scores. There was an outlier because one participant reported zero score for Pain Severity and Pain Interference. The findings for the individual physiological domain scores are presented in Table 9. There was a statistically significant, large positive correlation between Pain Severity and the frequency of physical symptoms reported (F-Physical). There was a statistically significant, small positive correlation between Pain Interference scores and F-Physical scores and moderately positive correlations between Pain Severity, Pain Interference, and Physical symptoms scores. Therefore, all variables within the physiological domain had a significant relationship with each other.

Table 9

Correlations of Physiological Sequelae (Pearson's Correlation) (n=93)

		Pain Severity	Pain Interference	Physical	F-Physical
Pain Severity	Pearson Correlation	1	**.462****	**.376****	**.504****
			(.000)	**(.000)**	**(.000)**
Pain Interference	Pearson Correlation	**.462****	1	**.438****	**.270****
		(.000)		**(.000)**	**(.009)**
Physical	Pearson Correlation	**.376****	**.438****	1	.006
		(.000)	**(.000)**		(.479)
F-Physical	Pearson Correlation	**.504****	**.270****	.006	1
		(.000)	**(.009)**	(.479)	

**. Correlation is significant at the 0.01 level (1-tailed). Probabilities in parentheses under correlations.

Note: Significant p-value in bold print

Psychological Sequelae. A Pearson's product-moment correlation was run to assess the relationship between the variables used for the psychological domain scores. There were no statistically significant correlations between the variables within the psychological domain. The frequency of psychological smptoms reported is denoted as F-Psychological. (See Table 10).

Table 10

Correlation of Psychological Sequelae (Pearson's Correlation) (n=93)

		Psychological	F-Psychological	BDI
Psychological	Pearson Correlation	1	.080	.036
			(.266)	(.370)
F-Psychological	Pearson Correlation	.080	1	-.056
		(.266)		(.333)
BDI	Pearson Correlation	.036	-.056	1
		(.370)	(.333)	

Probabilities in parentheses under correlations.

Relationship between Physiological Sequelae and Psychological Sequelae. A Pearson's product-moment correlation was run to assess the relationships between Pain Severity, Pain Interference, Physical, F-Physical, BDI, Psychological, and F-Psychological. There was a statistically significant small positive correlation between Pain Severity and F-Psychological. There were statistically significant small positive correlations between Pain Interference and Psychological. Physical had significant small positive relationships between Psychological, F-Psychological, and BDI. There were statistically significant correlations with F-Physical (See Table 11.)

Table 11

Correlations of Physiological Sequelae and Psychological Sequelae (Pearson's Correlation) (n=93)

Variables	Psychological	F-Psychological	BDI
Pain Severity	.115 (.141)	**.256*** **(.022)**	.076 (.241)
Pain Interference	**.224*** **(.016)**	.174 (.081)	.102 (.172)
Physical	**.279*** **(.004)**	**.200**** **(.054)**	**.177**** **(.048)**
F-Physical	-.002 (.494)	-.030 (.408)	.084 (.216)

* Correlations is significant at the 0.01 level (1-tailed).**Correlations is significant at the 0.05 level (1-tailed). Probabilities are in parentheses under the correlation.
Significant *p-values* in bold print.

Relationships between Physiological Sequelae and Sociocultural variables: Employment and/or School Involvement. A point-biserial correlation was run between Pain Severity, Pain Interference, Physical scores, frequency of physical symptoms (F-Physical), and the sociocultural variable (employment and/or school involvement). As previously mentioned, there were 39 (42%) participants who were not working or attending school and 53 (58%) participants who were socially involved by either working and/or attending school. All pain and physiological variables had a negative correlation with the sociocultural variable as predicted; however, only one was statistically significant. There was a statistically significant, negative correlation between the sociocultural variable and F-Physical score. Thus as the participants reported frequency of physical symptoms,

sociocultural involvement, i.e. employment and/or school attendance, decreased. (See Table 12.)

Table 12

Correlations between Physiological Sequelae and Sociocultural Variable: Employment and/or School Involvement (Pearson's Correlation) (n=93).

Variables	Sociocultural Variable (Employment/School Involvement)
Pain Severity	-.132 (.109)
Pain Interference	-.128 (.113)
Physical	-.152 (.074)
F-Physical	**-.205* (.025)**

Probabilities are noted in parentheses below correlation. *Correlation is significant at the 0.05 level (1-tailed).
Note: Significant *p-values* in bold print.

Relationship between Psychological Sequelae and Sociocultural Variables: Employment and School. A point-biserial correlation was run to determine the relationships between BDI scores, psychological symptom scores (Psychological), and frequency of reported psychological symptoms (F-Psychological), and the sociocultural variable i.e. employment and/or school involvement. There were no significant correlations between BDI, Psychological, F-Psychological, and Sociocultural. (See Table 13.)

Table 13

Correlations of Psychological Sequelae and Sociocultural Variables: Employment/School (Point-biserial Correlation) (n=93)

Variables	Sociocultural
Psychological	-.056 (.300)
F-Psychological	-.057 (.329)
BDI	.053 (.312)

Probabilities are noted in parentheses below correlation.

Categorical Relationships between Variables. A canonical correlation was performed between the sets of domain variables to determine their relationships with each other. The sets physiological sequelae domain variables and the psychological sequelae domain variables, were correlated with each other to assess their inter-relational characteristics as specified by the Chronic Disease Outcomes Triad (CDOT) Model. The physical set included the number and severity of physical symptoms often experienced by adults with SCD. The psychological set included the number and severity of mental/emotional symptoms often experienced by adults with SCD. The sociocultural variable was not included in this analysis because only one variable was significantly correlated with it, i.e., F-Physical and its relationship with other domains was apparent, based on the manner in which it was measured. The findings of the canonical correlations are provided in Table 14.

Table 14

Canonical Correlations and unstandardized coefficients between Physiological and Psychological domains and their corresponding canonical variates.

Canonical Correlations	First Canonical Variate Correlations	Coefficients	Second Canonical Variates Correlations	Coefficients	Third Canonical Variates Correlations	Coefficients
Physiological Sets						
Pain Severity	-.32	.08	-.77	-.24	.20	-.05
Pain Interference	-.67	-.04	-.38	.00	.62	.06
Physical	-.87	-.18	-.21	.06	-.44	-.15
F-Physical	-.21	-.17	-.25	.47	.48	.22
Psychological Sets						
Psychological	-.74	-.13	.14	.03	-.65	-.10
F-Psychological	-.25	-.10	-.97	-.38	.02	.03
BDI	-.59	-.08	.22	.02	.78	.09
Canonical Correlatons	.54		.31		.17	
Probabilities	***p* = .013**		*p* = .304			
Wilk's Lambda	.62		.07			

The set of physiological domain variables and psychological domain variables all significantly correlated with each other with a canonical variate correlation of .54, which had a significant probability of .013. The Wilk's Lambda was .62. Therefore, as the set of physiological domain variables increased, the set of psychological

domain variables also increased. The results support the stated hypothesis and the Chronic Disease Outcomes Triad (CDOT) Model.

Hypothesis 2*.* There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores with negative behavioral health outcome scores, and a negative relationship with positive behavioral health outcome scores; and, there will be a negative relationship of the sociocultural domain score with negative behavioral health outcome scores and a positive relationship with positive behavioral health outcome scores.

Spearman's rank-order correlations, Pearson's product-moment, and Point-biserial correlations were run to assess the relationship between the individual variables within the triadic sequelae with the variables representing behavioral health outcomes in adults with SCD. The Physical score was the only variable that statistically significantly correlated with all behavioral health outcomes in the direction as predicted. Therefore, as Physical scores increased, so did Fatalism and Perceived Prejudice scores, and Sickle Cell Self-Efficacy Scores (SCSES) decreased. There also were a statistically significant correlations in the predicted directions SCSES, Pain Severity scores, Pain Interference scores, and with the sociocultural variable (employment and/or school involvement). As Pain Severity and Pain Interference scores increased, so did employment and/or school involvement increase also. The BDI scores were significantly positively correlated with Fatalism and Perceived Prejudice scores in the predicted directions with all scores increasing or decreasing together. Hence, participants who report higher

depression were more likely to have higher fatalism and perceptions of prejudice toward them. (See Table 15.)

Table 15

Correlations of Triadic Sequelae with Behavioral Health Outcomes (Spearman's)

Variables	Fatalism	Perceived Prejudice	SCSES
Pain Severity	.130 (.114)	.118 (.138)	**-.343***** **(.0005)**
Pain Interference	-.052 (.315)	.148 (.084)	**-.266**** **(.006)**
Physical	**.221*** **(.019)**	**.209*** **(.025)**	**-.207*** **(.025)**
F-Physical	-.023 (.414)	.092 (.196)	-.090 (.199)
Psychological	-.002 (.495)	.034 (.375)	-.146 (.084)
F-Psychological	.087 (.254)	-.094 (.237)	-.131 (.158)
BDI	**.196*** **(.034)**	**.941***** **(.000)**	-.033 (.381)
Sociocultural	-.041 (.350)	-.058 (.296)	**.187*** **(.039)**

*. Correlation is significant at the 0.05 level (1-tailed).

**. Correlation is significant at the 0.01 level (1-tailed).

***Correlation is significant at the 0.001 level (1-tailed).

Note: Significant p-values in bold print. Probabilities are in parentheses under the correlation.

Hypothesis 3. There will be a positive relationship between negative behavioral health outcome scores (Fatalism and Perceived Prejudice) with each other, and an inverse relationship with positive behavioral health outcome scores (SCSES) in adults with SCD.

A Pearson's product moment was run to assess the relationships between Fatalism, Perceived Prejudice, and SCSES. There was a statistically significant, small positive relationship between Fatalism and Perceived Prejudice. Hence, when Fatalism perceptions increased Perceived Prejudice perceptions increased. There were no statistically significant correlations between Fatalism and SCSES and Perceived Prejudice and SCSES. (See Table 16.)

Table 16

Correlations between Behavioral Health Outcomes: Fatalism, Perceived Prejudice, and SCSES (Pearson's correlations) (n=93)

Variables	Fatalism	Perceived Prejudice	SCSES
Fatalism	1	**.223***	-.152
		(.019)	(.078)
Perceived Prejudice	**.223***	1	-.088
	(.019)		(.209)
SCSES	-.152	-.088	1
	(.078)	(.209)	

***.** Correlation is significant at the 0.05 level (1– tailed). Probabilities are in parentheses under the correlation. **Note: Significant *p*-values in bold print.**

Hypothesis 4. The number and severity of triadic domain scores will predict negative and positive behavioral health outcome scores.

Stepwise hierachal multiple regression analyses were conducted to determine the predictive ability of the triadic sequelae on the behavioral health

outcomes. There were separate analyses conducted for each behavioral health outcome, i.e., Fatalism, Perceived Prejudice, and SCSES. All variables were entered into each regression analyses in the following order; Model one Pain Severity, Pain Interference, Physical, and F-Physical, Model two added Psychological, F-Psychological, and BDI, and Model three added the sociocultural variable, employment and/or school involvement.

Although there was a small positive correlation between the variables Fatalism and Physical, there was no significant regression model. With the additions of each set of variables, the models slightly increased by 2%. Therefore, there was a weak predictive ability of the triadic sequelae with Fatalism as the dependent variable.

The following factors that significantly and positively correlated with Perceived Prejudice as the dependent variable were Pain Interference, Physical symptoms, and BDI scores. The addition of the psychological sequelae variables led to a statistically significant increase of the models by 78%. There was a slight increase when the sociocultural variable was added as a predictor. However, the predictive ability of the triadic sequelae and Perceived Prejudice as the dependent variable was weak; the means were far below the mean of the population.

The factors that significantly and negatively correlated with SCSES were Pain Severity, Pain Interference, and Physical symptoms. The factor that significantly positively correlated with SCSES was Sociocultural. The model with the physiological variables were significant, however with the addition of the psychological sequelae and sociocultural variables, the models were no longer

significant. The addition of psychological sequelae and sociocultural variables decreased the regression by 20%. These models had a strong predictive ability for the triadic sequelae and SCSES, the means were above the mean of the population. (See Tables 17-19 for the summaries of the regression analyses)

Table 17

Summary of heirarchal regression of triadic sequelae with Fatalism.

Fatalism Mean/SD 37.30/14.02	**B**	**SE-B**	***β***	***p-value***
Model 1				
Constant	18.73	10.38		.077
Pain Severity	.74	.45	.32	.106
Pain Interference	-.20	.15	-.21	.180
Physical	.83	.43	.28	.059
F-Physical	-1.06	1.31	-.14	.420
Model 2				
Constant	19.51	10.94		.081
Pain Severity	.72	.49	.31	.145
Pain Interference	-.20	.16	-.21	.203
Physical	.96	.50	.32	.062
F-Physical	-1.07	1.38	-.14	.442
Psychological	-.27	.34	-.12	.433
F-Psychological	.05	.75	.01	.943
BDI	.13	,24	.08	.592
Model 3				
Constant	24.43	12.14		**.050**
Pain Severity	.68	.49	.29	.172
Pain Interference	-.20	.16	-.21	.206
Physical	.84	.52	.28	.113
F-Physical	-1.09	1.38	-.14	.431
Psychological	-.24	.35	-.10	.501
F-Psychological	.05	.75	.01	.952
BDI	.15	.24	.09	.520
Sociocultural	-3.70	3.93	-.13	.352

R = Model 1 = .395, Model 2 = *.418*, Model 3 = *.435*. ΔR2= Model 1 = *.091*, Model 2 = *.057*, Model 3 = *.054*.
Significant *p-values* in bold print.

Table 18

Summary of Hierarchal regressions of triadic sequelae with Perceived Prejudice.

Perceived Prejudice Mean/SD 21.84/10.69	**B**	**SE-B**	***β***	***p-value***
Model 1				
Constant	5.88	7.89		.459
Pain Severity	-.53	.35	-.31	.132
Pain Interference	.15	.11	.22	.178
Physical	.54	.33	.24	.110
F-Physical	1.37	1.01	.23	.182
Model 2				
Constant	7.99	2.89		**.008**
Pain Severity	-.05	.13	-.03	.736
Pain Interference	-.03	.04	-.05	.437
Physical	.17	.13	.08	.210
F-Physical	.20	.37	.03	.586
Psychological	-.06	.09	-.03	.514
F-Psychological	-.15	.20	-.04	.471
BDI	1.18	.06	.94	**.000**
Model 3				
Constant	8.89	3.16		**.007**
Pain Severity	-.06	.13	-.03	.683
Pain Interference	-.03	.04	-.04	.461
Physical	.15	.13	.07	.279
F-Physical	.20	.37	.03	.594
Psychological	-.06	.09	-.03	.533
F-Psychological	-.14	.20	-.04	.483
BDI	1.19	.06	.94	**.000**
Sociocultural	-.75	1.03	-.04	.470

R = Model 1 = *.336*, Model 2 = *.946*, Model 3 = *.947*. ΔR2= Model 1 = *.046*, Model 2 = *.881*, Model 3 = *.880*.

Significant *p-values* in bold print.

Table 19

Summary of Hierarchal regressions of triadic sequelae with SCSES

Self-efficacy (SCSES) Mean/SD 33.93/5.60	**B**	**SE-B**	***β***	***p-values***
Model 1				
Constant	45.58	3.99		**.000**
Pain Severity	-.23	.18	-.25	.193
Pain Interference	-.00	.06	-.01	.955
Physical	-.35	.17	-.29	**.044**
F-Physical	-.02	.51	-.01	.966
Model 2				
Constant	45.89	4.20		**.000**
Pain Severity	-.20	.19	-.21	.303
Pain Interference	-.02	.06	-.05	.780
Physical	-.36	.19	-.30	.063
F-Physical	-.11	.54	-.03	.845
Psychological	-.03	.13	-.03	.811
F-Psychological	-.02	.29	-.01	.958
BDI	.09	.09	.14	.307
Model 3				
Constant	44.72	4.59		**.000**
Pain Severity	-.19	.19	-.20	.341
Pain Interference	-.02	.06	-.05	.757
Physical	-.34	.20	-.28	.091
F-Physical	-.10	.54	-.03	.853
Psychological	-.03	.13	-.04	.794
F-Psychological	-.02	.29	-.01	.947
BDI	.09	.09	.13	.350
Sociocultural	.98	1.49	.09	.516

R = Model 1 = *.458*, Model 2 = *.479*, Model 3 = *.486*. ΔR2= Model 1 = *.150*, Model 2 = *.122*, Model 3 = *.112*.
Significant *p-values* in bold print

Aim 2

To examine the relationship of demographic and disease-related characteristics of adults with SCD as predictors of triadic sequelae and behavioral health outcomes of SCD.

Hypothesis 5. The more frequently adults with SCD report the presence of diagnosed SCD complications, and hospitalizations/emergency department visits over the past year; the higher will be the number and severity of physiological and psychological domain scores, and the lower will be the sociocultural domain scores.

Pearson's product-moment and point-biserial correlations were run to assess the relationships between the presence of SCD Complications, the number of emergency room visits for pain crises in the past 12 months (PainER), the number of hospital admissions for pain crises in the past 12 months (PainHosp) and the triadic sequelae. There were statistically significant positive correlations between SCD Complications scores and Pain Severity scores, Pain Interference scores, Physical symptoms scores, and frequency of physical symptoms (F-Psychological) scores and a significant negative correlation with the sociocultural variable which is consistent with the hypothesized direction. Hence, when adults with SCD reported the presence of SCD Complications, physiological sequelae increased and the sociocultural variable decreased. In the analysis between SCD Complications and the sociocultural variable, there was an outlier based on a

participant who had 10 SCD Complications, who was not working or attending school.

Participants who reported increased visits to the ER for pain crises had an increase in Pain Severity and the frequency of physical symptoms and psychological symptoms. Adults with SCD who reported increase admissions to the hospital for pain crises also reported an increase in Pain Severity scores, Pain Interference scores, Physical symptoms scores, frequency of physical symptoms scores and the presence of psychological symptoms. (See Table 21 for correlation coefficients).

Table 20

Correlation Coefficients of Disease-related Characteristics and Triadic Sequelae. (Pearson's Correlation) (n = 93)

Variables	SCD Complications Correlation Coefficients	PainER Correlation Coefficients	PainHosp Correlation Coefficients
Pain Severity	**.265*** **(.006)**	**.224**** **(.018)**	**.171**** **(.055)**
Pain Interference	**.214**** **(.021)**	.140 (.093)	**.221**** **(.018)**
Physical	**.408*** **(.000)**	.069 (.258)	**.186**** **(.038)**
F-Physical	.107 (.155)	**.500*** **(.000)**	**.240**** **(.011)**
Psychological	.020 (.425)	-.010 (.462)	**.212**** **(.021)**
F-Psychological	**.247**** **(.009)**	**.184**** **(.040)**	-.113 (.189)
BDI	.073 (.284)	-.040 (.377)	.053 (.312)
Sociocultural	**-.227**** **(.015)**	.017 (.437)	-.024 (.411)

*Correlation is significant at a level of 0.001. **Correlation is significant at a level of 0.01. ***Correlation is significant at a level of 0.05. Probabilities are noted in parentheses below correlations.

Significant *p*-values are in bold print.

Hypothesis 6. Older age at diagnosis, older current age, and female gender will be positively associated with positive behavioral health outcomes (SCSES) and negatively associated with negative behavioral health outcomes (Fatalism and Perceived Prejudice).

Pearson's product-moment and point-biserial correlations were run to assess the relationships between Diagnosed Age, Current Age, Female gender and the behavioral health outcomes. There were 64 female participants in this study. There was a statistically significant positive correlation between Diagnosed Age and Perceived Prejudice which was not consistent with the hypothesized directions. However, female gender had a statistically negative correlation with Fatalism, which is consistent with the hypothesized direction. (See Table 22 for correlation coefficients).

Table 21

Correlations Coefficients of Characteristics of Participants and Behavioral Health Outcomes.

Variables	Fatalism	Perceived Prejudice	SCSES
Diagnosed Age	-.070	**.245***	-.022
	(.257)	**(.011)**	(.418)
Current Age	.042	.092	.087
	(.347)	(.195)	(.209)
Female	**-.168***	-.099	-.064
	(.057)	(.179)	(.276)

*Correlation is significant at the 0.05 level (1-tailed). Probabilities are in parentheses under correlation.
Note: Significant *p-values* in bold print.

Research Questions

Research Question 1. To what degree do the number and severity of physiological and psychological domain scores, and the sociocultural domain score predict diagnosed complications, and hospitalizations/emergency department visits over the past year?

Stepwise hierachal multiple regression analyses were conducted to determine the predictive ability of the triadic sequelae on the disease related characteristic. There were separate analyses conducted for each disease related characteristics SCD Complications, PainER, and PainHosp. All variables were entered into each regression analyses in the following order; Model one Pain Severity, Pain Interference, Physical, and F-Physical, Model two added Psychological, F-Psychological, and BDI, and Model three added Sociocultural.

The variables that were statistically significant and moderately positive with SCD Complications were Physical symptoms scores and Psychological symptoms scores. All regression models were significant however, when psychological sequelae and the sociocultural variable were added the model decreased from 24% to 6%. Physiological sequelae has a strong predictive ability of SCD Complications.

The variables that were statistically significant and positive with PainER were Pain Severity scores and the frequency of physical symptoms (F-Physical) scores. All regression models were significant and the strongest model included all the variables at 77% with 52% of the variance explained. Therefore, the triadic sequelae has a strong predictive ability of PainER.

There were no significant models between PainHosp and the triadic sequelae. Therefore, PainHosp was not a predictor of the triadic sequelae. (See Tables 22-24 for hierarchal summaries).

Table 22

Hierarchal summary of Triadic Sequelae with SCD Complications.

SCD Complications Means/SD 2.86/2.25	**B**	**SE-B**	***β***	***p-values***
Model 1				
Constant	-2.59	1.60		.102
Pain Severity	-.02	.07	-.06	.743
Pain Interference	-.00	.02	-.02	.884
Physical	.24	.07	.52	**.000**
F-Physical	.10	.20	.08	.632
Model 2				
Constant	-3.14	1.60		**.054**
Pain Severity	-.02	.07	-.04	.822
Pain Interference	-.00	.02	-.02	.883
Physical	.20	.07	.43	**.007***
F-Physical	.09	.20	.07	.651
Psychological	.09	.05	.24	.078
F-Psychological	.03	.11	.03	.812
BDI	-.03	.04	-.09	.478
Model 3				
Constant	-2.81	1.74		.113
Pain Severity	-.02	.07	-.05	.785
Pain Interference	-.00	.02	-.02	.903
Physical	.20	.07	.41	**.011***
F-Physical	.09	.21	.07	.658
Psychological	.09	.05	.24	.077
F-Psychological	.03	.11	.03	.805
BDI	-.02	.04	-.09	.521
Sociocultural	-.28	.57	-.06	.625

R = Model 1 = .490, Model 2 = .550, Model 3 = .553. ΔR^2 = Model 1 = .18, Model 2 = .21, Model 3 = .19. *Correlations is significant at 0.05 level (1-tailed).
Note: Significant *p-values* in bold print.

Table 23

Hierarchal summary of Triadic Sequelae with PainER.

PainER	**B**	**SE-B**	***β***	***p-values***
Model 1				
Constant	-1.36	.39		.**001**
Pain Severity	-.02	.02	-.16	.275
Pain Interference	-.01	.01	-.20	.107
Physical	.02	.02	.14	.211
F-Physical	.33	.05	.84	.**000**
Model 2				
Constant	-1.42	.41		**.001**
Pain Severity	-.01	.02	-.12	.472
Pain Interference	-.01	.01	-.22	.089
Physical	.01	.02	.07	.553
F-Physical	.32	.05	.82	**.000**
Psychological	.02	.01	.14	.216
F-Psychological	-.00	.03	-.02	.875
BDI	.00	.01	.03	.752
Model 3				
Constant	-1.82	.42		**.000**
Pain Severity	-.01	.02	-.08	.616
Pain Interference	-.01	.01	-.23	.059
Physical	.02	.02	.13	.279
F-Physical	.32	.05	.82	**.000**
Psychological	.01	.01	.13	.227
F-Psychological	-.01	.03	-.02	.828
BDI	.00	.01	.01	.961
Sociocultural	.33	.14	.24	**.019**

R = Model 1 = .72, Model 2 = .73, Model 3 = .77. ΔR^2 = Model 1 = .49, Model 2 = .47, Model 3 = .52. **Note: Significant *p-values* in bold print.**

Table 24

Hierarchal Summary of Triadic Sequelae with PainHosp.

PainHosp	B	SE-B	*β*	*p-values*
Model 1				
Constant	-.55	.53		.308
Pain Severity	-.01	.02	-.09	.673
Pain Interference	7.586E-5	.01	.00	.992
Physical	.02	.02	.11	.492
F-Physical	.13	.07	.34	.057
Model 2				
Constant	-.57	.55		.300
Pain Severity	.01	.03	.045	.832
Pain Interference	-.00	.01	-.04	.833
Physical	-.00	.03	-.02	.914
F-Physical	.10	.07	.26	.144
Psychological	.03	.02	.26	.079
F-Psychological	-.04	.04	-.14	.303
BDI	.01	.01	.07	.647
Model 3				
Constant	-.64	.60		.291
Pain Severity	.01	.03	.05	.811
Pain Interference	-.00	.01	-.04	.824
Physical	-.00	.03	-.01	.961
F-Physical	.10	.07	.26	.147
Psychological	.03	.02	.26	.083
F-Psychological	-.04	.04	-.14	.304
BDI	.01	.01	.06	.676
Sociocultural	.06	.19	.04	.776

R = Model 1 = .31, Model 2 = .41, Model 3 = .41. ΔR^2 = Model 1 = .02, Model 2 = .05, Model 3 = .03.

Research Question 2. To what degree do older age at diagnosis, older current age, and female gender predict behavioral health outcomes, (i.e., SCSES, Fatalism, and Perceived Prejudice)?

Stepwise hierarchal multiple regressions were conducted to analyze the predictive ability of the participant related characteristics on the behavioral health outcomes. There were separate analyses conducted for each participant related characteristic including Diagnosed Age, Current Age, and Female gender. All variables were entered into each regression analyses in the following order; Model one Diagnosed Age, Model two added Current Age, and Model three added Female gender.

There were no statistically significant variables with Fatalism and the participant related characteristics. There were no significant models and when all variables were added, the model provided only 20% of the prediction. Therefore, the participant related characteristics had a weak predictive ability on Fatalism.

The variable that was statistically significantly and positively correlated with Perceived Prejudice was Diagnosed Age. The model with Diagnosed Age was the only significant model. Models slightly increased by 6% when all variables were added. Therefore, participant related characteristics had a weak predictive ability of Perceived Prejudice.

There were no statistically significant variables with SCSES and the participant related characteristics in the model. Although, there were no significant models, the model increased by 11% when Current Age was added as a predictor but it decreased by 6% when Female gender was added to the prediction.

Therefore, the participant related characteristics had a weak predictive ability on SCSES. (See Table 25 for summaries of hierarchal regressions)

Table 25

Hierarchal Summary of Charateristics of Participants and Behavioral Health Outcomes.

Coefficients	B	SE-B	*B*	*p-values*
Fatalism				
37.30(14.02)				
Model 1				
Constant	39.14	3.16		**.000**
Diagnosed Age	-1.56	2.37	-.07	.513
Model 2				
Constant	37.05	4.26		**.000**
Diagnosed age	-2.35	2.61	-.11	.371
Current Age	.96	1.32	.09	.467
Model 3				
Constant	40.21	4.67		**.000**
Diagnosed Age	-2.28	2.59	-.10	.381
Current Age	1.05	1.31	.09	.424
Female	-5.09	3.19	-.17	.114
Perceived Prejudice				
21.84(10.69)				
Model 1				
Constant	16.96	2.35		**.000**
Diagnosed Age	4.14	1.76	.25	**.021**
Model 2				.
Constant	17.17	3.23		**.000**
Diagnosed Age	4.22	1.95	.25	**.033**
Current Age	-.10	.10	-.01	.922
Model 3				

Constant	18.76	3.57		**.000**
Diagnosed Age	4.29	1.95	.25	**.030**
Current Age	-.09	.10	-.01	**.**932
Female	-2.49	2.40	-.11	.302
SCSES 33.93(5.60)				
Model 1				
Constant	34.17	1.27		**.000**
Diagnosed Age	-.20	.95	-.02	.836
Model 2				
Constant	33.00	1.73		**.000**
Diagnosed Age	-.63	1.04	-.07	.550
Current Age	.53	.54	.12	.326
Model 3				
Constant	33.49	1.89		**.000**
Diagnosed Age	-.62	1.05	-.069	.559
Current Age	.55	.54	.12	.311
Female	-.82	1.28	-.07	.522

Fatalism-*R* = Model 1 = 0.07, Model 2 = 0.11, Model 3 = 0.20. ΔR^2 = Model 1 = -0.01, Model 2 = -0.01, Model 3 = 0.01.

Perceived Prejudice- *R* = Model 1 = 0.25, Model 2= 0.25, Model 3 = .27. ΔR^2 = Model 1 = 0.05, Model 2 = 0.04, Model 3 = 0.0

SCSES- *R* = Model 1 = 0.02, Model 2 = 0.11, Model 3 = 0.13. ΔR^2 = Model 1 = -0.01, Model 2 = -0.01, Model 3 = -0.02

Other Relevant Findings

Adults with SCD were very involved and invested to share their knowledge and experiences with this disease. The participants often made statements which were relevant to the study and recorded as notes by the investigator. Examples of statements that have relevance to the study's findings are as follows:

Some participants expressed feeling depressed from the effects of the COVID-19 pandemic because they were affected by the social shutdowns, loss of jobs, closure of schools, and experienced COVID-19 themselves. One participants expressed, "I feel depressed from the social isolation, I am a people person and now I'm sad because I can't be around people anymore." Several participants stated, "I was working until I got furloughed because of the pandemic" or " I contacted COVID and was hospitalized, and this took a lot out of me."

Some study participants shared stories of maltreatment by health care providers, family members, employers, and acquaintances. One study participant said, "I had to report a doctor to the hospital administration staff because of a racist remark he made while arguing with me about my treatment." Another study participant revealed, "I got a doctor fired from a hospital because of his racist attitude."

Several participants shared experiences of health care providers not believing they were in pain which led to under treatment and accusations of them being drug-seekers. One participant expressed, "I told the doctor in an emergency room that patients with SCD take pain meds daily and have been most of their

lives, so we developed a tolerance to the medications. Therefore, the average dosages or lower dosages of pain meds are not effective, we often require higher than normal dosages of opioids to alleviate pain that requires medical intervention." This statement shows how some SCD patients have to educate their health care providers and advocate for better treatment.

Some older participants were uncertain if they could keep sickle cell disease pain from interfering with their sleep or if they would be able to carry out activities of daily living. Some participants often expressed, "When my pain is an eight or higher, I need help getting dressed, thank God I have good support." An older participant who reported multiple physiological symptoms also noted that, "I am no longer sure if I can manage SCD symptoms as I get older."

Several participants expressed they would often wait until the pain was unbearable and their daily pain medication regimen was not effective before seeking medical interventions. They reported that it would often take days for them to decide to seek medical intervention. One study participant stated, "I won't go to the hospital unless my pain is an 8 or higher. I can handle anything under that."

As previously mentioned, many of the participants were employed and some had the responsibility of being parents, wives, and husbands; therefore, hospitalizations were very disruptive with their routine. One participant noted: "It's hard when I have to go to the hospital and leave my family. I have children but they understand that when mommy's sick she sometimes needs to go to the hospital to get better." Pain Interference was so prevalent in this sample that it caused great

disruption of their daily lives. One participant explained, “I would go to work in a lot of pain, but I hid it from my co-workers and friends because I didn’t want pity.” Another participant explained, “The pain is so unpredictable that this creates a major interference in my daily routine mainly because the pain crises can linger from two days to two weeks.” Another individual stated, “My crises were so severe that I had to retire early from teaching. I wouldn’t wish this pain on anyone.”

Several participants expressed their thoughts on death and dying. One participant stated, “We can’t help but think about dying because we were told by doctors all our lives we are going to die. First our parents are told, we won’t live past 10, then when I reached ten, we were told we won’t live past 14, then when I reached 14, they said we won’t live past 25, now I am 35 and still here.” Another participant expressed, “I often think about my own death when my friends with sickle cell disease die.”

There were insightful moments experienced by the researcher while interviewing the study participants. For example, several participants described self-taught distraction methods that often helped them cope with severe pain. One participant stated, “I paint when the pain is severe, this takes my mind off of the extreme pain.” Another individual stated, “I listen to music, it calms me down when I have a severe pain crisis.” Another participant described how health care providers display negative attitudes when they use their distraction methods and said, “I sit quietly and color to take my mind off the pain, then when I ask for pain meds, the nurse would say you’re not having pain”. Another insight gleaned was

how ambitious the participants were, despite experiencing debilitating and unpredictable SCD sequelae. For example, one noted, “I just started law school, it is already stressful, but I will keep going until I finish.” Another participant stated, “I own my own photography studio and I have many clients.” Another participant stated, “When the pandemic is over, I will resume my classes and get into medical school.” Such statements provide further evidence of the effects that sickle cell disease has on people living with this disease.

Summary

There were several statistically significant correlations within and between the variables of physiological sequelae, psychological sequelae, disease related and patient related characteristics. The variables representing the physiological sequelae were pain severity, pain interference, physical symptoms, and frequency of physical symptoms and several had moderately positive correlations with each other. The variables of the psychological sequelae were psychological, symptoms, frequency of psychological symptoms, and BDI depression scores which had few significant correlations with other variables. Physiological sequelae and psychological sequelae, had small positive correlations with pain severity and frequency of psychological symptoms and pain interference and psychological symptoms. Physical symptoms had significantly positive correlations with psychological symptoms, frequency of psychological symptoms, and BDI depression scores. The sociocultural variable, which measures participants’ social involvement in working and/or attending school, was found to have a small significant negative relationship with frequency of physical symptoms. The

physiological and psychological domain variable sets had significant and moderate correlations with each other using canonical correlations statistics. Pain severity, pain interference, and physical symptoms were found to have significant negative relationships with Sickle Cell Self-Efficacy scores and the sociocultural variable which represent employment and/or school involvement had a significantly positive relationship with Sickle Cell Self-Efficacy. Additionally, physical symptoms had small significant positive relationships with fatalism and perceived prejudice. BDI depression scores had a large positive relationship with preceived prejudice and a small positive relationship with fatalism**.**

There was a small positive relationship between fatalism and perceived prejudice in the sickle cell sample. The triadic sequelae had weak predictive ability related to the behavioral health outcomes. SCD Complications had small to moderate positive relationships with pain severity, pain interference, physical symptoms, and frequency of psychological symptoms and a small negative correlation with employment and/or school involvement. Pain that resulted in ER visits had a small positive relationship with pain severity scores and a large positive relationship with frequency of physical symptoms and frequency of psychological symptoms. Pain that resulted in hospitalization had a small positive relationships with pain severity, pain interference, physical symptoms, frequency of physical symptoms, and psychological symptoms. A small positive relationship was found between diagnosed age and perceived prejudice and a negative relationship was found between female gender and fatalism.

Physiological sequelae had a strong predictive ability on SCD Complications. The triadic sequelae had a strong predictive ability on pain that lead to ER visits and weak predictive ability on pain that resulted in hospitalizations. Patient related characteristics had weak predictive ability in relation to the behavioral health outcomes.

V. Discussion, Conclusions, and Recommendations

Discussion, Conclusions, and Recommendations

This predictive correlational study was developed to test the inter-relational and interactional relationships of variables derived based on the Chronic Disease Outcomes Triad (CDOT) Model as triadic sequelae of sickle cell disease (SCD) and as predictors of behavioral health outcomes in SCD. The interrelationships of SCD pain severity, pain interference, physical symptoms, frequency of physical symptoms, BDI depression, psychological symptoms, frequency of psychological symptoms, and employment and/or school involvement were examined in relation to behavioral health outcomes which included fatalism, perceived prejudice, and sickle cell self-efficacy. Additionally, other disease related and patient related characteristics such as the age at diagnosis, current age, pain episodes that led to emergency room (ER) visits, pain episodes that led to hospitalizations, SCD complications, and gender were examined in relation and predictive ability to fatalism, perceived prejudice, and sickle cell self-efficacy as behavioral health outcomes and the triadic sequelae of SCD. The Chronic Disease Outcomes Triad (CDOT) Model, which was developed by the P.I. based on relevant literature related to chronic diseases, was utilized as the conceptual framework that guided this study. This chapter presents a discussion of the study findings within the context of the CDOT Model, and examines the study's strengths and limitations. Implications and recommendations for clinical practice and future research are also provided.

Summary of the Research

The study sample of 93 adults with SCD was recruited either by email or the snowball method. Due to the COVID-19 pandemic and required Centers for Disease Control (CDC) guidelines, there were no face-to-face meetings for data collection; therefore, all interviews were conducted via telephone or online. The recruitment methods afforded a global population. The final sample was comprised of 64 female and 29 male adults between the ages of 18 to 74 years who were diagnosed with SCD. The variables under examination were categorized based on the CDOT Model as triadic sequelae into three domains: physiological sequelae, psychological sequelae, and sociocultural sequelae represented by one variable. The physiological sequelae were pain severity, pain interference, physical symptoms, and frequency of physical symptoms. The psychological sequelae were depression, psychological symptoms and frequency of psychological symptoms. The sociocultural variable was employment and/or school involvement. The behavioral health outcomes under examination included fatalism, perceived prejudice, and sickle cell self-efficacy. Other SCD related and patient related characteristics were age at diagnosis with SCD, which indicated the age the participant was diagnosed with SCD, current age, gender and additional variables addressed in the study were SCD complications, which indicated other SCD related illnesses, pain that led to ER visits, which indicated the number of times an individual visited the ER only for sickle cell pain episodes in the past 12 months prior to the study; and pain that led to hospitalization, which indicated the number of times an individual was admitted in the hospital for sickle cell pain

episodes during the past 12 months of the study. Table 26 summarizes the findings related to the study's hypotheses and research questions that were reported in Chapter IV.

Table 26

Summary of Significant Findings for each Hypothesis, Research Questions, and Additional Analyses.

Hypotheses/Research Questions	Significant Findings
Hypothesis 1: There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores, and a negative relationship of these two domains with the sociocultural domain scores when domain variables are assessed both individually and categorically.	Pain Severity, pain interference, physical symptoms, and frequency of physical symptoms were found to have significant positive correlations with each other. Physical symptoms and frequency of physical symptoms were not significantly correlated with each other. Frequency of psychological symptoms was not significantly correlated with Pain Interference and frequency of physical symptoms. Pain Severity and frequency of psychological symptoms had a significant positive correlation with each other. Pain interference and psychological symptoms were positively correlated. Physical symptoms were positively correlated with psychological symptoms, frequency of psychological symptoms, and depression. Employment and/or school involvement and frequency of physical symptoms had a significant negative relationship. Psychological symptoms, frequency of psychological symptoms, and depression scores were not significantly correlated with each other. The canonical correlation analyses revealed that the group of physiological domain variables were significantly and moderately correlated with the group of psychological domain variables. **Hypothesis 1 was partially supported by data analyses.**

Hypothesis 2: There will be a positive relationship between the number and severity of SCD physiological and psychological domain scores with negative behavioral health outcome scores, and a negative relationship with positive behavioral health outcome scores; and, there will be a negative relationship of the sociocultural domain score with negative behavioral health outcome scores and a positive relationship with positive behavioral health outcome scores.	Pain severity, pain interference, and physical symptoms had significant negative correlations with sickle cell self-efficacy. Employment and/or school involvement and sickle cell self-efficacy had a significant positive correlation. Physical symptoms had significant positive correlations with Fatalism and Perceived Prejudice. Pain severity, fatalism, and perceived prejudice had no significant correlations with each other. Depression scores, fatalism, and perceived prejudice had significant positive correlations with each other. Depression and sickle cell self-efficacy had no significant correlation with each other. Frequency of physical symptoms, psychological symptoms, and frequency of psychological symptoms had no significant correlations with any behavioral health outcomes. **Hypothesis 2 was partially supported by data analyses.**
Hypothesis 3: There will be a positive relationship between negative behavioral health outcome scores (sickle cell fatalism scores and perceived sickle cell prejudice scores) with each other, and an inverse relationship with positive behavioral health outcome	Perceived prejudice and fatalism were significantly and positively correlated as predicted. Sickle cell self-efficacy had no significant correlations with perceived prejudice scores, and fatalism scores.

scores (SCD self-efficacy scores) in adults with SCD.	**Hypothesis 3 was partially supported.**
***Hypothesis 4*:** The number and severity of triadic domain scores will predict negative and positive behavioral health outcome scores.	The strongest model for the prediction of fatalism included all variables of the triadic sequelae. The strongest models for the prediction of perceived prejudice were the models that added psychological, however, scores were below sample means. The strongest model for the prediction of sickle cell self-efficacy included all domain variables except for employment and/or school involvement which represented the sociocultural domain. **Hypothesis 4 was mostly supported by these analyses.**
Hypothesis 5: The more frequently adults with SCD report the presence of diagnosed SCD complications, and hospitalizations/emergency department visits over the past year; the higher will be the number and severity of physiological and psychological domain scores, and the lower will be the sociocultural domain scores.	SCD Complications had significantly positive correlations with pain severity, pain interference, physical, symptoms and frequency of psychological symptoms. It had a negative significant correlation with employment and/or school involvement as the sociocultural domain variable. Sickle cell pain that led to ER visits had significant positive correlations with pain severity frequency of physical symptoms and frequency of psychological symptoms. Sickle cell pain that led to hospitalizations had significant positive correlations with pain severity, pain interference, physical symptoms, frequency of physical symptoms and psychological symptoms. **Hypothesis 5 was mostly supported by data analyses.**

Hypothesis 6: Older age at diagnosis, older current age, and female gender will be positively associated with positive behavioral health outcomes (SCSES) and negatively associated with negative behavioral health outcomes (Fatalism and Perceived Prejudice).	Age at diagnosis with SCD and perceived prejudice had a significant positive correlation. Age at diagnosis, sickle cell self-efficacy, and fatalism had no significant correlation with each other. Current Age, fatalism, perceived prejudice, and sickle cell self-efficacy had no significant correlations with each other. Female gender and fatalism had a significantly negative correlation with each other. **Hypothesis 6 was partially supported by the data analyses.**
Research Question 1: To what degree do the number and severity of physiological and psychological domain scores, and the sociocultural domain score predict diagnosed complications, and hospitalizations/emergency department visits over the past year?	The regression model with the physiological sequelae had a strong ability to predict SCD Complications. The regression model that included all the domain variables had a strong predictive ability related to sickle cell pain that led to ER visits. None of the variables selected to represent the triadic sequelae were predictive of pain that led to hospitalizations. **Results for Research Question 1 partially supported the CDOT Model.**
Research Question 2: To what degree do older age at diagnosis, older current age, and female gender predict behavioral health outcomes, (i.e., SCSES, Fatalism, and Preceived Prejudice)?	The specified patient related characteristics had weak predictive ability related to fatalism, perceived prejudice, and sickle cell self-efficacy. **Results for Research Question 2 did not support the CDOT Model**

Discussion of Findings

A major underlying rationale for this study was to investigate the inter-relational and interactional relationship of the triadic sequelae in SCD as predictors of negative and/or positive behavioral health outcomes which is the basis of the Chronic Disease Outcomes Triad (CDOT) Model, which was developed by the PI for this study. The triadic sequelae, which includes physiological, psychological, and sociocultural sequelae are the attitudes, beliefs, symptoms and social circumstances that result from an individual having a disease and that affects their health outcomes. The study participants, which consisted of 93 adults with SCD, reported their experiences with sickle cell disease which served as the data for this study.

The Relationship of Triadic Sequelae Among Each Other in Sickle Cell Disease

Hypothesis 1 focused on determining how the number and severity of the sickle cell disease (SCD) physiological, psychological, and sociocultural sequelae impact adults with SCD between the ages of 18 to 65 years and older. As previously mentioned, it was postulated that the greater the number and severity of the sequelae, the more debilitating or confining they would be to the affected individual. In this population, the analyses supported hypothesis 1 and revealed pain severity, pain interference, number of physical symptoms, and frequency of physical symptoms all increased or decreased together and had significant relationships to each other. Therefore, the more a participant reported severe pain, they were more likely to report more interference of pain in their daily activities

and an increased number and frequency of other SCD physical symptoms. This is consistent with the literature that states that pain is the most prevalent and discomforting physiological sequelae (Anim, Osafo, & Young, 2016; Brown, et al., 2015; Barriteau & McNaull, 2018) which is most debilitating to those with SCD. Additionally, many individuals with SCD report that frequent, unpredictable occurrences of vaso-occlusive crises (VOCs) and other SCD symptoms are so severe that hospitalizations are required (USFDA, 2014), thus interfering with their functioning. The number of physical symptoms and the frequency of physical symptoms reported were not significantly associated with each other. Perhaps it is the nature of the physical symptom and the amount of discomfort or pain that is associated with it that is most important for those with SCD, rather than how frequently a symptom occurs.

Although depression was found among some of the study's participants, however there were no significant relationships with the other variables representing the psychological domain, i.e. number of psychological symptoms, and frequency of psychological symptoms. Depression is the most frequently cited psychological sequelae in SCD (Taylor, et al., 2013) and it has massive effects on the health-related quality of life of people living with SCD (Coleman, et al., 2016). However, most participants in this study reported somewhat low levels of depression, while also experiencing normal ups and downs. This may be related to the fact that interviews were conducted during the COVID-19 pandemic and some participants expressed feeling depressed from the effects of the pandemic.

There were participants who were affected by the social shutdowns, loss of jobs, closure of schools, and other negative aspects of experiencing COVID themselves.

Pain severity and frequency of psychological symptoms had a significantly positive relationship. Additionally, there was a significant positive relationship between pain interference, and psychological symptoms, and physical symptoms with the number of psychological symptoms, frequency of psychological symptoms, and depression. These findings are consistent with prior research with persons with SCD which revealed that there is a relationship between pain intensity and negative moods (Taylor et al., 2013). Other research revealed recurrent and frequent episodes of VOCs may be associated with psychosocial problems such as reduced participation in normal activity of daily living, as well as with depression, and anxiety; therefore, cognitive therapy should be considered as a nonpharmacologic option for pain management (Uwaezuoke, et al., 2018). When pain is at its maximum it interrupts their ability to sleep which can lead to feelings of sadness, anger, worry, and nervousness. The more the participants reported pain interfering with different aspects of their lives both physically and psychologically, such as their general activity, their mood, their ability to walk, their relationship with other people, weakness, pain, and swelling of hands and feet they will more likely to experience psychological symptoms such as feeling sad, tense or nervous, short-tempered, worried or concerned, and depression. Additionally, participants who reported the presence of pain, weakness, swelling of hands or feet, and shortness of breath, they were more likely to experience increased

frequency of feeling sad, feeling worried or concerned, feeling tense or nervous, and depression.

There was a significantly negative relationship between the frequency of physical symptoms and employment and/or school involvement. Suggesting that frequent reports of physical symptoms such as the presence of pain, interference with sleep, and interference with one's ability to walk or engage in other physical activities can negatively impact the ability of those with SCD to work and/or attend school. Involvement in work and/or school was not significantly associated with any of the other psychological or physiological domain variables. Therefore, higher frequency of physical symptoms appears to be a dominant determinant of ability to work and/or attend school in adults with SCD.

When variables in the physiological and psychological domains were correlated with each other and analyzed categorically, the group of variables in the physiological domain was moderately correlated with the group of variables in the psychological domain. This supports the inter-relationship between physical and psychological domains as postulated by the CDOT Model.

The Association of Triadic Sequelae with Behavioral Health Outcomes

According to Strickland (2013), behavioral health outcomes of a chronic disease are how one acts, thinks, or behaves due to the consequences of a disease. Behavior health outcomes may be positive or negative, which may affect one's self-care, coping strategies, and motivation for self-care and adaptive responses related to the disease (Strickland, 2013). Fatalistic beliefs stem from a

high degree of suffering from the disease, negative attitudes and belief about the disease, and the beliefs that individuals are powerless to influence their health or illness because these are controlled by external forces (Strickland, 2001; Ramona & Carmona, 2018). Perceived chronic disease prejudice stems from the belief that others view persons with a specific chronic disease negatively because they have the disease (Carlisle, 2015). Additionally, perceived prejudice as a chronic stressor tends to produce negative outcomes (Carlisle, 2015; Haywood, et al., 2014). Self-efficacy was a positive behavior health outcome and is demonstrated when a person takes charge of life situations and has a strong expectation that they will be successful with disease outcomes (Adegbola, 2011).

As postulated based on the CDOT Model, the participants' data in this study revealed a significant positive correlation between the two negative behavioral outcome variables, i.e. fatalism and perceived prejudice; and nonsignificant negative correlations between theses variables and sickle cell self-efficacy, the positive outcome variable. As suggested by the CDOT Model, significant negative relationships were found between pain severity, pain interference, and physical symptoms in relation to SCD sickle cell self-efficacy. On the other hand, SCD self-efficacy was significantly positively associated with employment and/or school involvement, the variable that represented the sociocultural domain in this study. This indicated that individuals who reported increased severe pain, pain interference, and the presence of physical symptoms reported less certainty in their ability to control their SCD and its health outcomes. However, those with more certainty of their ability to control their disease were more likely to work and/or

attend school. This is consistent with the literature which reported that lower levels of self-efficacy were associated with more symptoms, higher pain severity, and frequent physician visits (Matthie, Jenerette, & McMillan, 2015; Matthie, et al., 2019). Also, this study revealed that physical symptoms were significantly and positively associated with fatalism, and perceived prejudice. Therefore, participants reporting the presence of physical symptoms also believed they could not effectively influence their health care because of external forces, e.g. negative interactions they have encountered in the past, or feeling discriminated against because of their SCD. For example, prior research revealed that patients' perceptions of a dispute with healthcare workers was attributed to both their race/ethnic status and the fact that they had SCD (Haywood, et al., 2014). Often, study participants shared stories of maltreatment from health care providers, family members, employers, and acquaintances.

As postulated, depression was significantly and positively associated with fatalism and perceived prejudice. Individuals in this study who reported higher levels of depression also reported higher levels of perceived prejudice and fatalistic beliefs. This is consistent with the research that revealed depressive symptoms are closely related to the social devaluation that adults with SCD experience known as stigma (Holloway, McGill, & Bediako, 2016). These findings also are consistent with research that revealed perceived prejudice about a disease may result in even greater susceptibility to negative chronic disease outcomes (Carlisle, 2015). Additionally, other research revealed a large amount of chronic disease fatalism is likely to lead to negative emotional responses (Strickland, et al., 2001).

Predictors of Behavioral Health Outcomes from the Triadic Sequelae

Based on statistical analyses that focused on predictors of the behavioral outcomes (fatalism, perceived prejudice, and sickle cell self-efficacy) in this study, the strongest predictors of fatalism was a combination of all variables in this study that represented the triadic sequelae. This is consistent with a recent study on fatalism and diabetes that revealed disease fatalism is described is an emotional frustration resulting from lifestyle disruptions from managing the conditions resulting from the disease (Abbott, Slate, Graven, Lemecks, & Grant, 2021). Indeed, the variables in this study that represented the physiological, psychological, and sociocultural domains measure factors that could be expected to disrupt a person's hopefulness and outlook related to having sickle cell disease.

The strongest predictive model for perceived prejudice was found when the psychological sequelae were included as the set of predictors. Given that perceived prejudice is based on emotional responses and negative perceptions of behaviors of others associated with their disease, this finding is not surprising. This is consistent with the literature that shows persons who are preoccupied with sickle cell prejudice are more likely to be characterized by negative emotions such as depression, anxiety, anger, and feelings of hopelessness (Strickland, et al., 2001; Maddray & Phillips, 2020). This is consistent with research that revealed many health care professionals often under treat the painful sickle cell episodes because of their negative attitudes towards patients with SCD and their suspicion

that these patients have a drug addiction (Institute of Medicine, 2012; Freiermuth et al., 2014; Brennan-Cook et al., 2018; Maddray & Phillips, 2020).

The strongest prediction model for sickle cell self-efficacy resulted when all variables of the triadic sequelae were included as predictors. In this study, the more the participants reported severe pain, interference of pain, and the presence and frequency of physical symptoms, the more they reported lower levels of self-efficacy. Again, this is consistent with literature that reveals lower levels of self-efficacy were correlated with high pain severity (Matthie, Jenerette, & McMillan, 2015; Molter & Abrahamson, 2015).

Demographic and Disease-Related Characteristics and their Relationships with Triadic Sequelae and Behavioral Health Outcomes

This study sought to assess the association of demographic and disease-related characteristics, SCD Complications, sickle cell pain episodes that led to ER visits, and sickle cell pain episodes that led to hospitalizations with the triadic sequelae. In this study, SCD Complications consisted of the diagnosis of other disease-related complications such as stroke, avascular necrosis (AVN), lung problems, eye problems, and painful and swollen joints. Many participants had at least two SCD Complications which may lead to other difficulties related to living with SCD such as problems ambulating, poor cognition, and limited ability to care for oneself (Frostholm et al., 2018). Additionally, these complications often occur after multiple experiences with vaso-occlusive pain crises that resulted in long-term organ damage caused by repeated ischemia of the organ tissues (Coleman et al., 2016; Frostholm et al., 2018). As expected, based on the CDOT Model, as

SCD complications increased, so also did pain severity, pain interference, physical symptoms, and frequency of psychological symptoms. Therefore, these participants reported increased pain severity, increased pain interference, presence of more physical symptoms and more frequent psychological symptoms when SCD complications increased. It is not surprising that participants with increased SCD Complications had less involvement in employment and/or school attendance since such complications often reduce one's ability to function physically.

Twelve percent of this study's population reported sickle cell pain episodes that led to ER visits five times or more within the past 12 months prior to the study. As sickle cell pain episodes that led to ER visits increased, so also did pain severity, frequency of physical symptoms, and the frequency of psychological symptoms. As sickle cell pain episodes that led to hospitalizations increased, pain severity, pain interference, the number of physical symptoms, and psychological symptoms reported also increased. This population's daily medication treatment regimen included long-acting and short-acting opiates and NSAIDs. However, when pain is so severe and the daily treatment regimens are not effective, the affected individuals can be expected to visit the emergency room (ER) for aggressive analgesic management. This explains why pain severity, frequency of physical symptoms, and sickle cell pain episodes that led to ER visits had positive relationships.

A rather large percent (13%) of the study's participants reported sickle cell pain that led to hospitalization five times or more during the past 12 months of the study. These results highlight the potential physical and emotional toll and burden that sickle cell pain episodes can place on those affected. Such findings are consistent with the analyses which revealed significant positive relationships between sickle cell pain that led to hospitalizations, pain severity, pain interference, frequency of physical symptoms, and presence of psychological symptoms. Findings related to age at diagnosis, current age, and female gender were assessed in relation to their association with behavioral health outcomes. Age at diagnosis and perceived prejudice were significantly and positively associated with each other. It is possible that older age at diagnosis helped participants to more readily notice and label other's negative behaviors toward the disease as more prejudicial. Age at diagnosis is important to consider within this study because prior research shows that the earlier an individual experiences the onset of signs and symptoms the more likely will the disease and its consequences be more severe (Archer, Galacteros, & Brugnara, 2015; Jonassaint et al., 2016; Azar & Wong, 2017). As participants experience more disease symptoms, they are more likely to exhibit negative behavioral health outcomes (Strickland et al., 2001). Gender and fatalism had a negative relationship, which indicated that estimated effects were driven by the results for males. In other words, in this study more males had fatalistic beliefs than females. Additionally, current age had no significant relationships and female gender did not have significant relationships with perceived prejudice and sickle cell self-efficacy.

This study also sought to determine the relative contribution of the triadic sequelae to disease related characteristics, SCD Complications, sickle cell pain episodes that led to ER visits, and sickle cell pain that led to hospitalizations. As previously noted, variables of the triadic sequelae were entered sequentially to determine which variables had the strongest predictive ability of each disease related characteristic. It was not surprising that the strongest predictive variables for SCD Complication were the physiological domain variables only because most of the complications represent complications of physiological changes due to sickling of red blood cells that result in ischemia of body organs.

The strongest predictive variables for sickle cell pain episodes that led to ER visits included all variables in the study that represented the triadic sequelae. However, the triadic sequelae had insignificant predictive ability of sickle cell pain episodes that led to hospitalizations. This may be due to statistical artifact, i.e. the small number of participants (low variance) who reported hospitalizations due to sickle cell pain. The demographic related characteristics had some predictive ability on the behavioral health outcomes.

Conclusions

Sickle cell disease is one of the most debilitating chronic diseases with symptoms that are unpredictable and often difficult to control. This hereditary hematologic disease can affect almost every organ of the body due to multiple repeated and lifelong VOCs that result in both acute and chronic pain. Adults with SCD often have severe physiological involvement, such as pain severity and

frequent physical symptoms, which often begin early childhood. In addition to the extensive physiological damage caused by the disease, these patients experience psychological sequelae and negative sociocultural effects. The CDOT Model proposes that physiological sequelae, psychological sequelae, and sociocultural sequelae of the disease have an inter-relational and interactional relationship with each other, and have an impact on the affected person's behavioral health outcomes. The results of this study supported most of the key propositions of the CDOT Model and that the triadic sequelae were inter-relational and interactional.

Therefore, conclusions of this study include the following:

1. Adults with SCD who report severe pain are more likely to also experience an increase of pain interference in their lives and daily activities, as well as an increase in the number and severity of sickle cell physical symptoms, such as their ability to walk and engage in other important life activities, such as work or attending school.

2. When adults with SCD have frequent and severe pain and physical symptoms, pain and symptoms are more likely to interfere with different aspects of their lives, but persons with SCD also are impacted psychologically and may experience depression. They also will experience frequent psychological symptoms such as feeling sad, are more likely to be tense or nervous, short-tempered, worried or concerned.

3. Adults with SCD who report the presence of physical symptoms such as weakness, yellowing of skin or eyes, or eye problems are also more likely to have more psychological symptoms.

4. Adults with SCD who have increases in severe pain, interference with daily activities from pain, and increased presence of physical symptoms are more likely to have less self-efficacy and will be less certain of their ability to control their health outcomes.

5. Adults with SCD with multiple and more severe symptoms are more likely to perceive that they have little influence over their health care because of their perceptions of discrimination from others due to their SCD.

6. Adults with SCD who reported high levels of depression are also more likely to perceive that they are being discriminated against and have fatalistic beliefs because of their SCD.

7. Adults with SCD who have greater perceptions of perceived prejudice by others against SCD, also will more likely have fatalistic perceptions about SCD.

8. The triadic sequelae moderately predicted fatalistic attitudes in adults with SCD. The psychological domain moderately predicted perceived prejudice. The physiological and psychological sequelae of SCD are predictive of self-efficacy in adults with SCD.

9. When adults with SCD report increased pain severity, pain interference, presence of physical symptoms, and frequency of psychological symptoms they

more likely to also report a diagnosis of at least one other SCD complication such as stroke, AVN, or eye problems.

10. Adults who visited the ER five times or more for sickle cell pain episodes within the past 12 months are also more likely to report increased pain severity and frequency of physical symptoms.

11. Adults who were admitted in the hospital for sickle cell pain episodes within the past 12 months of the study also reported increased pain severity, pain interference, presence of more physical symptoms, frequency of physical symptoms, and the presence of more psychological symptoms.

12. The earlier a person was diagnosed with SCD, they more likely they were to have experienced symptoms of SCD at an early age.

13. Physiological sequelae in SCD strongly predict the presence of SCD Complications. The physiological and psychological sequelae and involvement in work and/or school, a sociocultural sequelae in this study are predictive of sickle cell pain episodes that lead to ER visits but not predictive of sickle cell pain episodes that lead to hospitalizations.

14. Demographic related characteristics were not predictive of the behavioral health outcomes (fatalism, perceived prejudice, and SCD self-efficacy) that were the focus of this study.

15. The results of this study partially supported the propositions of the CDOT Model developed by the PI.

Implications of Findings for Clinical Practice

As noted by participants' statements above, adults with SCD often must live with the debilitating, unpredictable, and difficult to control effects of this disease. Throughout the course of their lives, many have to live with pain which can disrupt their daily routines, and they sometimes encounter unfriendly health care establishments and personnel. They fight to be heard and confront death of people with SCD close to them. This study provided quantitative and qualitative documentation of their perceptions of the prejudice and emotional suffering they perceived as they live with their condition.

Often when persons with SCD encounter the health care system, they are in a critical, vulnerable state. However, some health care providers know very little about the patient's suffering beyond the painful episodes and crises which are reflected in their perceptions of discrimination and prejudice of others related to their disease as noted by Strickland and associates (2001). The findings of one or two studies do not change the clinical practice circumstances. However, the distinctive findings of this study, in a field where there is limited research of this kind, are a significant contribution to the limited evidence of how the inter-related and interactional triadic sequelae can impact behavioral health outcomes.

Most importantly, healthcare providers should not only familiarize themselves with pathophysiological effects of this life-long disease but also be aware of the psychological and sociocultural effects this disease may have on the behavioral outcomes of persons with SCD. These effects can be caused by the

pathophysiological aspects of the disease; however social stigma and negative attitudes from health care providers can ultimately influence the person's behavioral health outcomes. Furthermore, health care providers need to know that most adults with SCD, will likely visit the hospital or ER when the pain is too severe to manage themselves at home, or when psychological symptoms are present, and when there are other complications. Healthcare providers can gain great knowledge on how to individualize the treatment and management of this disease, by listening to the patient.

The CDOT Model, investigated the physiological, psychological, and sociocultural effects as an inter-relational and interactional relationship of sequelae of adults with SCD. The results of this study partially supported the propositions of the CDOT Model suggesting that adults with SCD often experience sequelae which may be debilitating and devastating to the persons with SCD. Using the CDOT Model as a basis for assessing patients' needs and developing treatment plans could provide a more effective treatment approach and a more appropriate perspective when caring for adults with SCD.

Implications for Future Research

This study was informative about adults with SCD and the physiological, psychological, and sociocultural effects or sequelae of this disease. It is the first to explore this issue from this approach, since the CDOT Model was developed to guide the development and testing of the study hypotheses. The CDOT Model can also be used to guide the study of other chronic diseases and further elucidate the theorectical underpinnings of chronic diseases and chronic disease care. The CDOT Model devised for this study is a tool that can be considered as a conceptual framework for qualitative studies also. The propositions of the CDOT Model may be used to investigate the inter-relational and interactional relationships of the triadic sequelae and its impact on behavioral outcomes with other chronic diseases such as fibromyalgia, diabetes, and renal failure.

Future research on this topic is necessary and should include the CDOT Model to guide the identification of physiological, psychological, and sociocultural effects of sequelae of disease, adequate treatment approaches, and management of the sequelae of SCD and other chronic diseases. Patient's perceptions of death and dying and the impact of treatment issues, such as the opioid crisis in SCD, and how it may interfere with adequate treatment are needed. More sociocultural factors that impact health outcomes, such as spirituality, religiosity, and social support need to be more carefully investigated.

Transitional adults with SCD, that is adults between the ages of 18 years to 25 years old, who are transitioning from pediatric health care to adult care

should be studied more intensely to assess how their transition experiences within health care facilities are related to their physiological, psychological, and sociocultural sequelae. This study found that males with SCD had more fatalistic attitudes, Further exploration of the attitudes of males with SCD and their behavioral health outcomes would be useful in elucidating correlates of fatalism and other behavioral health outcomes. Given the state of race relations in America and the fact that SCD is more common in Blacks, consideration should be given to determining the relationship between perceived racism and physiological, psychological, and sociocultural sequelae in SCD.

Study participants were entrepreneurs, community organizers, attending medical school, attending law school, and practicing law; however, these participants reported pain severity, pain interference, and a wide range of number and severity of physiological and psychological symptoms. The quantitative findings provided in this study of self-taught distraction methods for controlling and coping with SCD pain, deserve further investigation as possible treatment approaches.

Strengths of the Study

This study has several unique strengths. First, this study provided data of the inter-relational and interactional effects of a chronic disease on the behavioral health outcomes of adults with SCD. This is the first study that investigated physiological, psychological, and sociocultural sequelae in one study. Second, the study includes the perception of SCD from participants globally. Third, this study

addresses the role that chronic disease triadic sequelae play in helping to mold behavioral health outcomes which is important for determining what can be expected when caring for persons with SCD. Fourth, this study provided a platform for adults with SCD to share their perceptions of living with SCD in a safe, non-judgmental environment. Another strength of this study is the information obtained regarding sickle cell disease and how it effects the whole person. This study provided an opportunity for persons living with this disease to gain further knowledge of their mental/emotional state as it relates to SCD. Lastly, this study was the first to implement the theoretical propositions of the CDOT Model.

Limitations of the Study

Several limitations are present and should be noted in this study. The interviews began at the beginning of the COVID-19 pandemic which interfered with several aspects of the study. The shutdown of face-to-face interactions due to the COVID-19 pandemic guidelines limited contact between people due to social distancing guidelines. As a result, there were no in-person doctor visits, support group meetings, and conferences therefore, the researcher had to recruit participants via email or telephone. This allowed the researcher to reach the population that would have attended support groups and conferences by telephone and the internet via zoom. The COVID-19 pandemic was an on-going pandemic which resulted from coronavirus 2 (SARS-CoV-2) which caused by severe acute respiratory syndrome in those affected by it. After being identified in December 2019, the World Health Organization (WHO) declared an international public health emergency on January 30, 2020 and then a pandemic on March 11, 2020. Millions

of Americans were affected by the disease and to date, there are over 500,000 deaths in America related to COVID-19.

This pandemic has resulted in significant global social and economic disruption which led to the full or partial closure of educational institutions, public areas, and businesses. Hospitals became overrun by COVID-19 patients, and all Americans were asked to wear masks whenever they encountered others. Furthermore, the pandemic had a large effect on participants of this study including some participants contacting the virus or losing a loved-one to the virus.

It is possible that depression, loss of employment, and not attending school due to the pandemic could have influenced the results of the study. For example, levels of depression reported could have been impacted by the pandemic. Therefore, the degree to which the COVID-19 pandemic impacted the data collected for this study cannot be determined. from the pandemic which was not determined in this study. This study was limited because it only addressed one sociocultural variable within the sociocultural domain. More salient sociocultural variables, such as spirituality and social support, should be integrated into future research with persons with SCD while using the CDOT Model Other salient variables within the physiological and psychological domains should also be investigated. Another limitation of this study was the sample size, more participants could have afforded more significant results.

All the measures utilized in this study were self-report. A reliance on self-report measures open the possibility of inaccuracy in reporting and the possibility

of poor recall. Other more biological-based variables and measures should be integrated into the design of the study.

Summary

This predictive correlational study was developed to examine the inter-relational and interactional relationship of the triadic sequelae in SCD as predictors of SCD behavioral health outcomes in adults with SCD. The CDOT Model was developed by the researcher to further investigate chronic diseases and common behavioral health outcomes frequently affiliated with chronic diseases. This study was the first research to test the theoretical propositions of this conceptual model. The perceptions of adults with SCD were explored and offered vast knowledge on how they are shaped from the physical, psychological, and sociocultural effects of the disease.

Findings partially supported the CDOT Model and provided insights into the influencing factors in the lives of adults with SCD. Results indicated the physical, mental/emotional, and social involvement has an effect on the perceptions of living with this disease. A significant finding was the number and severity of the physiological sequelae, psychological sequelae, and sociocultural sequelae influences both negative and positive behavioral health outcomes. Furthermore, the presence of other complications is significantly and positively related to the physiological and psychological domain variables and negatively related to employment and/or school involvement. When physiological and psychological sequelae are severe and frequent, this leads to increased ER and hospitalizations.

Males had a significant relationship with fatalistic beliefs more than females. The older age at diagnosis had significantly and positively relationships with perceived prejudice and fatalism. Lastly, pain is a constant element in their lives and sometimes the pain may interfere with their daily activities but this study revealed how they are able to adjust their lives and live to its fullest.

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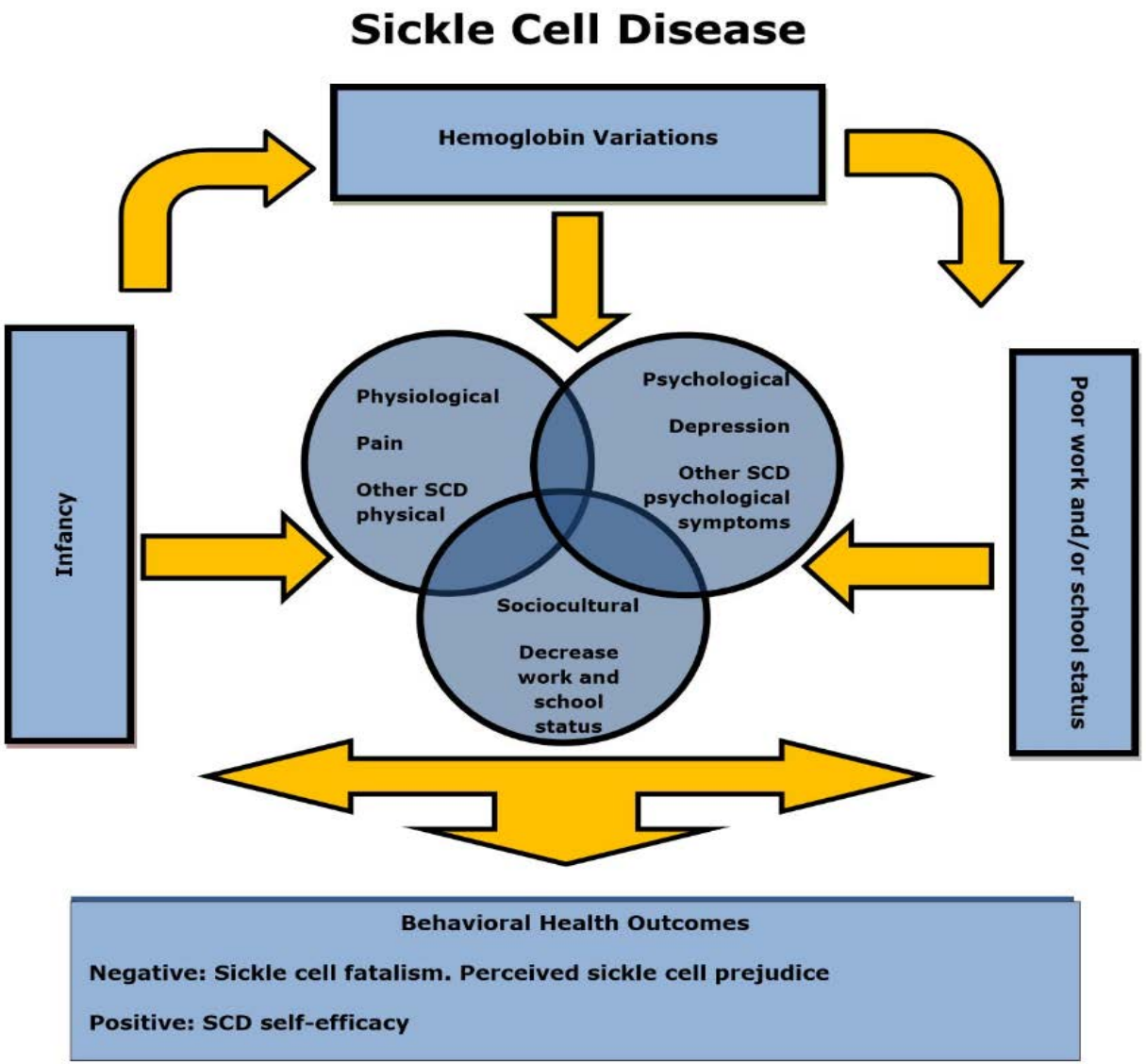
Appendices

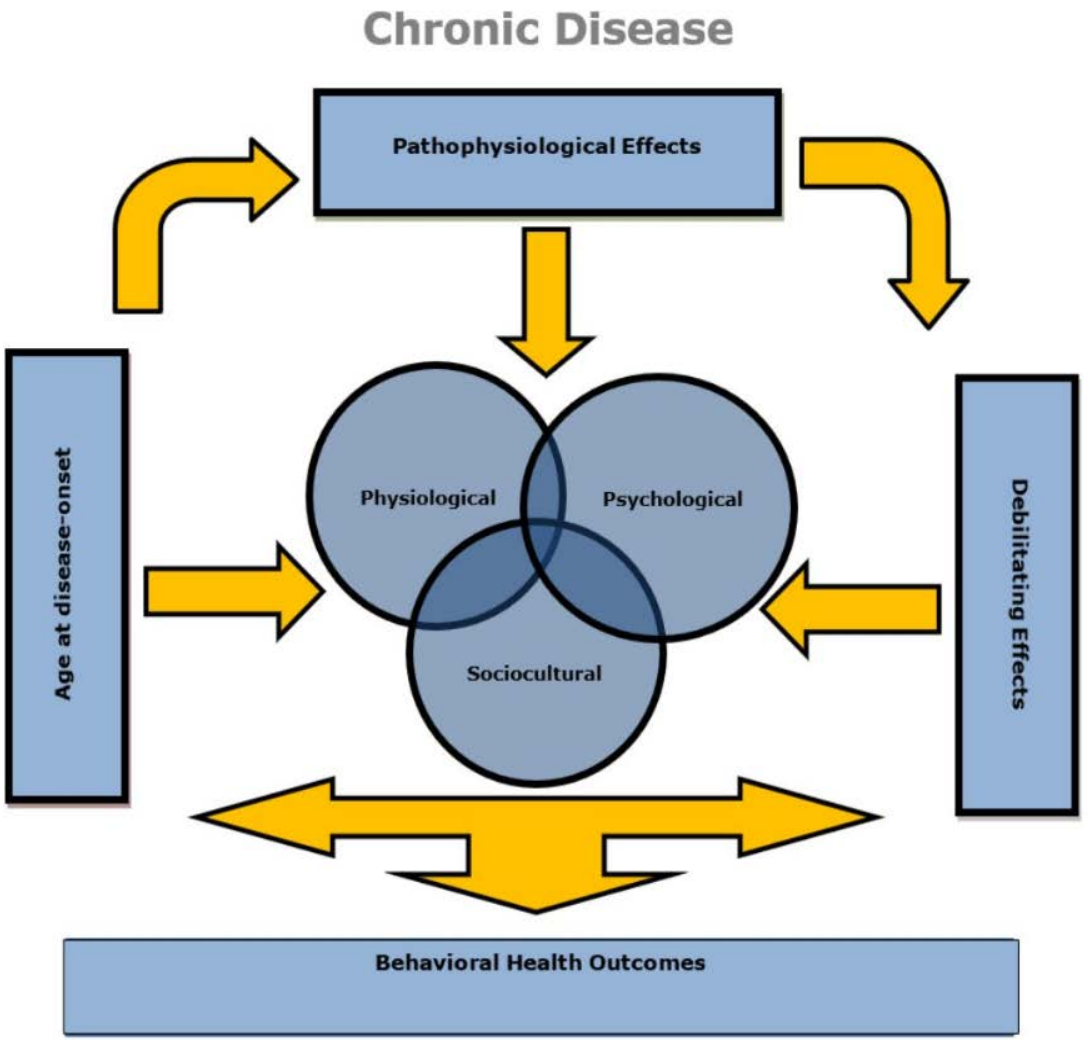
Appendix A

Conceptual Model for Sickle Cell Disease


Sickle Cell Disease
Hemoglobin Variations
Infancy
Physiological
Pain
Other SCD physical
Psychological
Depression
Other SCD psychological symptoms
Sociocultural
Decrease work and school status
Poor work and/or school status
Behavioral Health Outcomes
Negative: Sickle cell fatalism. Perceived sickle cell prejudice
Positive: SCD self-efficacy

Appendix B

Conceptual Framework Model


Chronic Disease
Pathophysiological Effects
Age at disease-onset
Physiological
Psychological
Sociocultural
Debilitating Effects
Behavioral Health Outcomes

Appendix C

Study Advertisement Flyer

FIU Nicole Wertheim College of Nursing & Health Sciences
FLORIDA INTERNATIONAL UNIVERSITY

SICKLE CELL DISEASE PATIENTS NEEDED

If you are 18 years old and older and have been diagnosed with sickle cell disease or sickle cell anemia, then please help with this survey.

What is the survey about?

◊ A study to assess how sickle cell disease affects your life physically, mentally, and socially.

What is involved?

⇒ Participate in telephone interview or complete online questionnaire less than 45 minutes to complete.
⇒ Names will not be used and all information is confidential.
⇒ There are no right or wrong answers and you may stop at any time.
⇒ You can receive a $10.00 e-gift card.

How can you help?

⇒ Agree to participate
⇒ Be willing to share your opinions about living with sickle cell disease.

Who is eligible?

⇒ Adult men and women with a diagnosis of sickle cell disease
⇒ NOT in active pain crises
⇒ Have NOT previously participated in this study.

For more information contact :
Lisa Gay Fryar, Ph.D. candidate, MSN, RN
Tel. (305) 606-1220 **Email:** lwood006@fiu.edu ***Or***
Ora Lea Strickland, PhD, DSc(Hon), RN, FAAN,
Nicole Wertheim College of Nursing & Health Sciences,
Florida International University, **Tel.** (305) 348-0407
Email: olstrick@fiu.edu

Appendix D

Chronic Disease Attitude Scale

Chronic Disease Attitude Scale

DIRECTIONS: Below are a number of statements about sickle cell disease. Look at the words at each end of the scale below each statement. For each statement, *circle the number on the scale from 0 to 10 that best shows how you feel*. Remember, there is no right or wrong answers. Whatever you think is the right answer.

In the scales below "0" means you completely disagree, "5" means that you do not really agree or disagree and 10 means that you totally agree with the statement.

Please respond to each of the following statements based on what you believe.

1. If someone has a chronic disease it was meant to be.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately agree — Agree

2. If someone has a chronic disease that's the way they were meant to die.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately agree — Agree

3. A person with a chronic disease should expect to live a long life.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

4. If someone has symptoms of a chronic disease it doesn't matter what they do, they will have them anyway.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

5. If someone has a chronic disease it is already too late to do anything about it.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

6. If someone has a chronic disease it doesn't matter if they go to the doctor/health care provider, they will get sick anyway.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

7. Having a chronic disease makes people think about dying a lot.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

8. If someone has a chronic disease, medications/treatments won't make any difference.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

9. If someone has a chronic disease they will never be able to take care of themselves.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

10. If someone has a chronic disease they will have problems in their personal relationships.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

11. If someone has a chronic disease people believe they can do well in their job.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

12. People believe it is hard to have fun with someone who has a chronic disease.

0 1 2 3 4 5 6 7 8 9 10

Disagree — Moderately Agree — Agree

13. People with a chronic disease are fine unless they are having symptoms.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

14. People do not want to be in a relationship with a person who has a chronic disease.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

15. People with a chronic disease use their condition to get attention.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

16. People with a chronic disease use their condition to get drugs or treatments they do not need.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

17. People with a chronic disease will have difficulty raising children.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

18. People who have a chronic disease cannot be good employees.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

19. If someone has a chronic disease they will not have a productive life.

0 1 2 3 4 5 6 7 8 9 10

Disagree Moderately Agree Agree

Appendix E

Consent Form and Survey Questionnaire

Chronic Disease Outcomes Triad

Start of Block: Default Question Block

ADULT VERBAL CONSENT TO PARTICIPATE IN A RESEARCH STUDY
The prediction of behavioral health outcomes in adults with sickle cell disease using the Chronic Disease Outcomes Triad Model

SUMMARY INFORMATION

Things you should know about this study:
Purpose: The purpose of the study is to assess physical, mental, and social impact of living with sickle cell disease. **Procedures**: If you choose to participate, you will be asked to provide your name and contact number or email address to me only. Answer questions and share your experiences over the phone or online (If any question makes you uncomfortable, you may skip it). Questions will include topics such as personal questions including age, ethnic background, education level as well as questions about how sickle cell disease may be affecting your mental and emotional health. If your responses to some of these questions indicate that you may be at risk for elevated depression the researcher will follow-up with you via telephone and provide you with some referrals for additional support. **Duration:** This will take about will take between 45 to 60 minutes of your time via a telephone interview with me. **Risks**: There are questions that may reveal emotional consequences based on responses. **Benefits:** There are no foreseeable benefits to participate in this study. **Alternatives:** There are no known alternatives available to you other than not taking part in this study. **Participation:** Taking part in this research project is voluntary.
Please carefully read the entire document before agreeing to participate.

PURPOSE OF THE STUDY

The purpose of this study is to assess how physical, mental, and social

symptoms associated with sickle cell disease affect your beliefs, perceptions, and management of this condition.

NUMBER OF STUDY PARTICIPANTS

If you decide to be in this study, you will be one of 120 people in this research study.

DURATION OF THE STUDY

Your participation will take 45 to 60 minutes of your time. If you agree to be in the study, I will ask you to do the following things:

1. Provide your name and contact number to the researcher only. Email address will be required for participants who desire to answer survey online.

2. Answer the questions and share your opinions over the phone or submit online.

RISKS AND/OR DISCOMFORTS

Potential for injury from participation in this study is minimal. Completion of telephone interviews or on-line surveys are free of risks for physical harm. However, some questions may raise personal issues that could be emotionally upsetting.

BENEFITS

There is no direct benefit from this study beyond self-sacrificing rewards for participating in the development of science in this topic area. There are no direct potential benefits to society except the gaining of knowledge about a population of patients living with sickle cell disease.

ALTERNATIVES

There are no known alternatives available to you other than not taking part in this study.

CONFIDENTIALITY

The records of this study will be kept private and will be protected to the fullest extent provided by law. In any sort of report, we might publish, we will not include any information that will make it possible to identify you. Research records will be stored securely, and only the researcher team Lisa G. Fryar and Dr. O. Strickland will have access to the records. However, your records may be inspected by authorized University or other agents who will also keep the information confidential.

USE OF YOUR INFORMATION

Identifiers about you will be removed from the identifiable private information and that, after such removal, the information could be used for future research studies or distributed to another investigator for future research studies without additional informed consent from you or your legally authorized representative.

COMPENSATION & COSTS

There is no compensation for participating in this study. There are no costs to you for participating in this study.

RIGHT TO DECLINE OR WITHDRAW

Your participation in this study is voluntary. You are free to participate in the study or withdraw your consent at any time during the study. You will not lose any benefits if you decide not to participate or if you quit the study early. The investigator reserves the right to remove you without your consent at such time that he/she feels it is in the best interest.

RESEARCHER CONTACT INFORMATION

If you have any questions about the purpose, procedures, or any other issues relating to this research study you may contact Lisa G. Fryar at 305-606-1220, email: lwood006@fiu.edu or Dr. O Strickland at FIU Nicole Wertheim College of Nursing and Health Sciences at 305-348-0231.

IRB CONTACT INFORMATION

If you would like to talk with someone about your rights of being a subject in this research study or about ethical issues with this research study, you may contact the FIU Office of Research Integrity by phone at 305-348-2494 or by email at ori@fiu.edu .

Do you provide your consent to participate in this research project?

O Yes, I consent to participate in this research project. (4)

O No, I do not consent (6)

Click to write the question text

O Email Address (4)

_____

O Country where you live (5)

_____

The following questions will assess your physical symptoms while living with sickle cell disease.

Q1 Throughout our lives, most of us have had pain from time to time (such as minor headaches, sprains, and toothaches). Have you had pain other than these everyday kinds of pain today?

- O Yes (1)
- O No (2)

Q2 Where on your body do you feel pain?
Select all that apply

- ☐ head (1)
- ☐ neck (2)
- ☐ shoulders (3)
- ☐ elbows (4)
- ☐ wrists (5)
- ☐ hands (6)
- ☐ fingers (7)
- ☐ chest (8)

- [ ] stomach (9)
- [ ] genitals/groin (10)
- [ ] hips (11)
- [ ] knees (12)
- [ ] ankles (13)
- [ ] feet (14)
- [ ] toes (15)
- [ ] upper back (16)
- [ ] middle back (17)
- [ ] lower back (18)
- [ ] buttocks (19)
- [ ] shins (20)

- [ ] thighs (21)
- [ ] arms (22)
- [ ] legs (23)
- [ ] face (24)

Q3 What area hurts the most?
Select all that apply:

- [ ] head (1)
- [ ] neck (2)
- [ ] shoulders (3)
- [ ] elbows (4)
- [ ] wrists (5)
- [ ] hands (6)
- [ ] fingers (7)
- [ ] chest (8)

- [ ] stomach (9)
- [ ] genitals/groin (10)
- [ ] hips (11)
- [ ] knees (12)
- [ ] ankles (13)
- [ ] feet (14)
- [ ] toes (15)
- [ ] upper back (16)
- [ ] middle back (17)
- [ ] lower back (18)
- [ ] buttocks (19)
- [ ] thighs (20)

- [ ] arms (21)
- [ ] legs (22)
- [ ] shins (23)
- [ ] face (24)

Q4 Please rate your pain by marking the box beside the number that best describe your pain at its WORST in the last 30 days.

- O 0- No pain (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)

O 8 (9)

O 9 (10)

O 10- Pain as bad as you can imagine (11)

Q5 Please rate your pain by marking the box beside the number that best describes your pain at its LEAST in the last 30 days.

O 0- No pain (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

O 5 (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Pain as bad as you can imagine (11)

Q6 Please rate your pain by marking the box beside the number that best describes your pain on the average

O 0- No pain (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

O 5 (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Pain as bad as you can imagine (11)

Q7 Please rate your pain by marking the box beside the number that tells how much pain you have right now.

- O 0- No pain (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Pain as bad as you can imagine (11)

Q8 What treatments or medications are you receiving for your pain?
Please type in answers

_____
___

_____
___

_____
___

_____
___

_____
___

Q9 In the last 30 days, how much relief have pain treatments or medications provided? Please mark the box beside the percentage that most shows how much RELIEF you have received.

- O 0 %- No relief (1)
- O 10% (2)
- O 20% (3)
- O 30% (4)
- O 40% (5)
- O 50% (6)
- O 60% (7)
- O 70% (8)
- O 80% (9)
- O 90% (10)
- O 100%- Complete relief (11)

Q10 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your general activity.

- O 0- Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q11 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your mood.

- O 0- Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q12 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your walking ability

- O 0- Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q13 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your normal work (work outside the home and housework) and/or school attendance.

- O 0- Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q14 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your relations with other people.

- O 0- Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q15 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your sleep

- O 0-Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q16 Mark the box below the number that describes how, during the past 30 days, pain has interfered with your enjoyment of life.

- O 0-Does not interfere (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5 (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Completely interferes (11)

Q17 Rate the frequency of the following symptoms over the preceding 6 months: weakness

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q18 Rate the frequency of the following symptoms over the preceding 6 months: yellowing of skin or eyes

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q19 Rate the frequency of the following symptoms over the preceding 6 months: vomiting

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q20 Rate the frequency of the following symptoms over the preceding 6 months: nausea

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q21 Rate the frequency of the following symptoms over the preceding 6 months: pain

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q22 Rate the frequency of the following symptoms over the preceding 6 months: heart problems

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q23 Rate the frequency of the following symptoms over the preceding 6 months: gall stones

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or more times) (3)
- O 4 = very often (six or more times) (4)

Q24 Rate the frequency of the following symptoms over the preceding 6 months: eye trouble

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or more times) (3)
- O 4 = very often (six or more times) (4)

Q25 Rate the frequency of the following symptoms over the preceding 6 months: kidney problems

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or more times) (3)
- O 4 = very often (six or more times) (4)

Q26 Rate the frequency of the following symptoms over the preceding 6 months: swelling of hands or feet

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or more times) (3)
- O 4 = very often (six or more times) (4)

Q27 Rate the frequency of the following symptoms over the preceding 6 months: shortness of breath

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or more times) (3)
- O 4 = very often (six or more times) (4)

Thank you for providing your responses to the physical symptoms questions related to living with sickle cell disease.The next set of questions will assess the mental / emotional impact that you may experience while living with sickle cell disease.

Q28 Please choose the best response:

- O 0 = I do not feel sad (1)
- O 1 = I feel sad (2)
- O 2 = I am sad all the time and I can't snap out of it (3)
- O 3 = I am so sad and unhappy that i can't stand it (4)

Q29 Please choose the best response:

- O 0 = I am not particularly discouraged about the future (1)
- O 1 = I feel discouraged about the future (2)
- O 2 = I feel I have nothing to look forward to (3)
- O 3 = I feel the future is hopeless and that things cannot improve (4)

Q30 Please choose the best response:

- O 0 = I do not feel like a failure (1)
- O 1 = I feel I have failed more than the average person (2)
- O 2 = As I look back on my life, all I can see is a lot of failures (3)
- O 3 = I feel I am a complete failure as a person (4)

Q31 Please choose the best response:

- O 0 = I get as much satisfaction out of things as I used to (1)
- O 1 = I don't enjoy things the way I used to (2)
- O 2 = I don't get real satisfaction out of anything anymore (3)
- O 3 = I am dissatisfied or bored with everything (4)

Q32 Please choose the best response:

- O 0 = I don't feel particularly guilty (1)
- O 1 = I feel guilty a good part of the time (2)
- O 2 = I feel quite guilty most of the time (3)
- O 3 = I feel guilty all of the time (4)

Q33 Please choose the best response:

- O 0 = I don't feel I am being punished (1)
- O 1 = I feel I may be punished (2)
- O 2 = I expect to be punished (3)
- O 3 = I feel I am being punished (4)

Q34 Please choose the best response:

- O 0 = I don't feel disappointed in myself (1)
- O 1 = I am disappointed in myself (2)
- O 2 = I am disgusted with myself (3)
- O 3 = I hate myself (4)

Q35 Please choose the best response:

- O 0 = I don't feel I am any worse than anybody else (1)
- O 1 = I am critical of myself for my weaknesses or mistakes (2)
- O 2 = I blame myself all the time for my faults (3)
- O 3 = I blame myself for everything bad that happens (4)

Q36 Please choose the best response:

- O 0 = I don't have any thoughts of killing myself (1)
- O 1 = I have thoughts of killing myself, but I would not carry them out (2)
- O 2 = I would like to kill myself (3)
- O 3 = I would kill myself if I had the chance (4)

Q37 Please choose the best response:

- O 0 = I don't cry any more than usual (1)
- O 1 = I cry more now than I used to (2)
- O 2 = I cry all the time now (3)
- O 3 = I used to be able to cry, but now I can't cry even though I want to (4)

Q38 Please choose the best response:

- O 0 = I am no more irritated by things than I ever was (1)
- O 1 = I am slightly more irritated now than usual (2)
- O 2 = I am quite annoyed or irritated a good deal of the time (3)
- O 3 = I feel irritated all the time (4)

Q39 Please choose the best response:

- O 0 = I have not lost interest in other people (1)
- O 1 = I am less interested in other people than I used to be (2)
- O 2 = I have lost most of my interest in other people (3)
- O 3 = I have lost all my interest in other people (4)

Q40 Please choose the best response:

- O 0 = I make decisions (1)
- O 1 = I put off making decisions more than I used to (2)
- O 2 = I have greater difficulty in making decisions more than I used to (3)
- O 3 = I can't make decisions at all anymore (4)

Q41 Please choose the best response:

- O 0 = I don't feel that I look any worse than I used to (1)
- O 1 = I am worried that I am looking old or unattractive (2)
- O 2 = I feel there are permanent changes in my appearance that make me look unattractive (3)
- O 3 = I believe that I look ugly (4)

Q42 Please choose the best response:

- O 0 = I can work about as well as before (1)
- O 1 = It takes an extra effort to get started at doing something (2)
- O 2 = I have to push myself very hard to do anything (3)
- O 3 = I can't do any work at all (4)

Q43 Please choose the best response:

- O 0 = I can sleep as well as usual (1)
- O 1 = I don't sleep as well as I used to (2)
- O 2 = I wake up 1-2 hours earlier than usual and find it hard to get back to sleep (3)
- O 3 = I wake up several hours earlier than I used to and cannot get back to sleep (4)

Q44 Please choose the best response:

- O 0 = I don't get more tired than usual (1)
- O 1 = I get tired more easily than I used to (2)
- O 2 = I get tired from doing almost anything (3)
- O 3 = I am too tired to do anything (4)

Q45 Please choose the best response:

- O 0 = My appetite is no worse than usual (1)
- O 1 = My appetite is not as good as it used to be (2)
- O 2 = My appetite is much worse now (3)
- O 3 = I have no appetite at all anymore (4)

Q46 Please choose the best response:

- O 0 = I haven't lost much weight, if any, lately (1)
- O 1 = I have lost more than five pounds (2)
- O 2 = I have lost more than ten pounds (3)
- O 3 = I have lost more than fifteen pounds (4)

Q47 Please choose the best response:

O 0 = I am no more worried about my health than usual (1)

O 1 = I am worried about physical problems like aches, pains, upset stomach, or constipation (2)

O 2 = I am very worried about physical problems and its hard to think of much else (3)

O 3 = I am worried about my physical problems that I cannot think of anything else (4)

Q48 Please choose the best response:

O 0 = I have not noticed any recent change in my interest in sex (1)

O 1 = I am less interested in sex than I used to be (2)

O 2 = I have almost no interest in sex (3)

O 3 = I have lost interest in sex completely (4)

Q49 Rate the frequency of the following psychological symptom over the preceding 6 months: feeling sad

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q50 Rate the frequency of the following psychological symptom over the preceding 6 months: feeling tense or nervous

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q51 Rate the frequency of the following psychological symptom over the preceding 6 months: feeling short-tempered

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q52 Rate the frequency of the following psychological symptom over the preceding 6 months: feeling worried or concerned

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q53 Rate the frequency of the following psychological symptom over the preceding 6 months: problems coping

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q54 Rate the frequency of the following psychological symptom over the preceding 6 months: problems sleeping

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q55 Rate the frequency of the following psychological symptom over the preceding 6 months: problems eating

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

Q56 Rate the frequency of the following psychological symptom over the preceding 6 months: problems paying attention

- O 1 = never or rarely (zero or one time) (1)
- O 2 = not very often (two or three times) (2)
- O 3 = often (four or five times) (3)
- O 4 = very often (six or more times) (4)

" Believe you can and you are half way there"
- Theodore Roosevelt

Thank you for sharing your mental / emotional health responses while living with sickle cell disease.

The next set of questions aim to better understand the individual living with sickle cell disease.

Q57 Gender

- [ ] Male (1)
- [ ] Female (2)

Q58 Age

- O 18-24 (1)
- O 25-34 (2)
- O 35-44 (3)
- O 45-54 (4)
- O 55-64 (5)
- O 65 or more (6)

Q59 Ethnic Origin

- O Black or African-American (1)
- O Asian/Pacific Islander (2)
- O Hispanic or Latino (3)
- O Native American or American Indian (4)

- O White (5)
- O Other (6)

Q60 Education

- O Less than HS diploma (1)
- O High School (2)
- O Some college (3)
- O Bachelor's degree (4)
- O Graduate degree (5)

Q61 Insurance coverage

- O No insurance (1)
- O Private insurance (2)
- O Medicaid/Obamacare (3)
- O Medicare (4)
- O Government (5)

Q62 What genotype of SCD do you have?

- O HbSS (1)
- O HbS/b-O thalassemia (2)
- O HbS/b+ thalessemia (3)
- O HbSC (4)
- O HbSD (5)
- O HbSE (6)
- O HbSS-thalessemia (7)

Q63 Relationship Status

- O Single (1)
- O Divorced (2)
- O Married (3)
- O In a relationship (4)

Q64 Are you currently working?

- O Yes (1)

O No (2)

Q65 Are you currently attending school?

O Yes (1)

O No (2)

Q66 How old were you when you were diagnosed with SCD?

O 0-7 years (1)

O 8-16 years (2)

O 16-24 years (3)

O 24 years or more (4)

Q67 When was your last sickle cell pain crisis?

O Less than 1 week ago (1)

O 1-2 weeks ago (2)

O 2-4 weeks ago (3)

O 1-3 months ago (4)

O 4-6 months ago (5)

O 7-9 months ago (6)

O 10-12 months ago (7)

O 1 year ago or more (8)

Q68 How many times in the past 12 months were you treated for sickle cell pain crisis in emergency room only?

O 0-1 (1)

O 2-4 (2)

O 5 or more (3)

Q69 How many times in the past 12 months were you admitted to a hospital for treatment of sickle cell pain crises?

O 0-1 (1)

O 2-4 (2)

O 5 or more (3)

Q70 Were you diagnosed with other SCD complication? If so, please select all that apply

☐ Stroke (1)

- [ ] Enlarged liver (2)
- [ ] Kidney disease (3)
- [ ] Painful and swollen joints (4)
- [ ] Leg sores (5)
- [ ] Heart problems (6)
- [ ] Lung complication (7)
- [ ] Pulmonary Hypertension (8)
- [ ] Avascular Necrosis (AVN) (9)
- [ ] Restless Legs Syndrome (10)
- [ ] Priapism (11)
- [ ] Mental Illness (12)
- [ ] Eye Problems (13)

The final set of questions will assess your perception of living with sickle cell disease.

Q72 If someone has sickle cell disease it was meant to be.

- O 0-Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5-Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q73 If someone has sickle cell disease that's the way they were meant to die.

- O 0-Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5-Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q74 A person with sickle cell disease should expect to live a long life.

- O 0- Disagree (1)

- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q75 If someone has symptoms of sickle cell disease it doesn't matter what they do, they will have them anyway.

- O 0-Disagree (1)
- O 1 (2)
- O 2 (3)

- O 3 (4)
- O 4 (5)
- O 5-Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q76 If someone has sickle cell disease it is already too late to do anything about it.

- O 0-Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)

- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q77 If someone has sickle cell disease it doesn't matter if they go to the doctor/health care provider, they will get sick anyway.

- O 0-Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5-Moderately agree (6)
- O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10-Agree (11)

Q78 Having sickle cell disease makes people think about dying a lot.

O 0-Disagree (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

O 5-Moderately agree (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Agree (11)

Q79 If someone has sickle cell disease, medications/treatments won't make any difference.

O 0- Disagree (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

O 5- Moderately agree (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Agree (11)

Q80 If someone has sickle cell disease, they will never be able to take care of themselves.

- O 0- Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q81 If someone has sickle cell disease, they will have problems in their personal relationship.

- O 0- Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5-Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q82 If someone has sickle cell disease people believe they can do well in their jobs.

- O 0- Disagree (1)

- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

"Nothing is IMPOSSIBLE. The word itself says I'M POSSIBLE!"
- Audrey Hepburn

Q83 People believe it is hard to have fun with someone who has sickle cell disease.

- O 0- Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q84 People with sickle cell disease are fine unless they are having symptoms.

- O 0- Disagree (1)

- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q85 People do not want to be in a relationship with a person who has sickle cell disease.

- O 0- Disagree (1)
- O 1 (2)
- O 2 (3)

O 3 (4)

O 4 (5)

O 5- Moderately agree (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Agree (11)

Q86 People with sickle cell disease use their condition to get attention.

O 0- Disagree (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q87 People with sickle cell disease use their condition to get drugs or treatments they do not need.

- O 0- Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Agree (11)

Q88 People with sickle cell disease will have difficulty raising children.

O 0- Disagree (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

O 5- Moderately agree (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Agree (11)

Q89 People who have sickle cell disease cannot be good employees.

O 0- Disagree (1)

O 1 (2)

O 2 (3)

O 3 (4)

O 4 (5)

O 5- Moderately agree (6)

O 6 (7)

O 7 (8)

O 8 (9)

O 9 (10)

O 10- Agree (11)

Q90 If someone has sickle cell disease they will NOT have a productive life.

- O 0- Disagree (1)
- O 1 (2)
- O 2 (3)
- O 3 (4)
- O 4 (5)
- O 5- Moderately agree (6)
- O 6 (7)
- O 7 (8)
- O 8 (9)
- O 9 (10)
- O 10- Agree (11)

Q91 How sure are you that you can do something to cut down on most of the pain you have when having a pain episode?

- O Not sure at all (1)

O Not sure (2)

O Neither (3)

O Sure (4)

O Very sure (5)

Q92 How sure are you that you can keep doing most of the things you do day-to-day?

O Not at all sure (1)

O Not sure (2)

O Neither (3)

O Sure (4)

O Very sure (5)

Q93 How sure are you that you can keep sickle cell disease pain from interfering with your sleep?

O Not sure at all (1)

O Not sure (2)

O Neither (3)

O Sure (4)

O Very sure (5)

Q94 How sure are you that you can reduce your sickle cell disease pain by using methods other than taking extra medication?

O Not sure at all (1)

O Not sure (2)

O Neither (3)

O Sure (4)

O Very sure (5)

Q95 How sure are you that you can control how often or when you get tired?

- O Not sure at all (1)
- O Not sure (2)
- O Neither (3)
- O Sure (4)
- O Very sure (5)

Q96 How sure are you that you can do something to help yourself feel better if you are feeling sad or blue?

- O Not sure at all (1)
- O Not sure (2)
- O Neither (3)
- O Sure (4)
- O Very sure (5)

Q97 As compared with other people with sickle cell disease, how sure are you that you can manage your life from day-to-day?

- O Not sure at all (1)
- O Not sure (2)
- O Neither (3)
- O Sure (4)
- O Very sure (5)

Q98 How sure are you that you can manage your sickle cell disease symptoms so that you can do the things you enjoy doing?

- O Not sure at all (1)
- O Not sure (2)
- O Neither (3)
- O Sure (4)
- O Very sure (5)

Q99 How sure are you that you can deal with the frustration of having sickle cell disease?

- O Not sure at all (1)
- O Not sure (2)
- O Neither (3)
- O Sure (4)
- O Very sure (5)

Appendix F

Florida InternationalUniversity
Internal Review Board Approval

FLORIDA INTERNATIONAL UNIVERSITY

Office of Research Integrity
Research Compliance, MARC 414

MEMORANDUM

To:	Dr. Ora Lea Strickland
CC:	Lisa Fryar
From:	Maria Melendez-Vargas, MIBA, IRB Coordinator
Date:	April 3, 2020
Protocol Title:	"The Prediction of Behavioral Health Outcomes in Adults with Sickle Cell Disease using the Chronic Disease Outcomes Triad Model."

The Health Sciences Institutional Review Board of Florida International University has approved your study for the use of human subjects via the **Expedited Review** process. Your study was found to be in compliance with this institution's Federal Wide Assurance (00000060).

IRB Protocol Approval #:	IRB-20-0123	**IRB Approval Date:**	04/03/20
TOPAZ Reference #:	108839	**IRB Expiration Date:**	04/03/23

As a requirement of IRB Approval you are required to:

1) Submit an IRB Amendment Form for all proposed additions or changes in the procedures involving human subjects. All additions and changes must be reviewed and approved by the IRB prior to implementation.
2) Promptly submit an IRB Event Report Form for every serious or unusual or unanticipated adverse event, problems with the rights or welfare of the human subjects, and/or deviations from the approved protocol.
3) Utilize copies of the date stamped consent document(s) for obtaining consent from subjects (unless waived by the IRB). Signed consent documents must be retained for at least three years after the completion of the study.
4) **Obtain continuing review and re-approval of the study prior to the IRB expiration date**. Submit the IRB Renewal Form at least 30 days in advance of the study's expiration date.
5) Submit an IRB Project Completion Report Form when the study is finished or discontinued.

HIPAA Privacy Rule: N/A

Special Conditions: N/A

For further information, you may visit the IRB website at http://research.fiu.edu/irb.

MMV/em

Appendix G

IRB Approval Letter Amendment

FIU | FLORIDA INTERNATIONAL UNIVERSITY

Office of Research Integrity
Research Compliance, MARC 414

MEMORANDUM

To:	Dr. Ora Lea Strickland
CC:	Lisa Fryar
From:	Maria Melendez-Vargas, MIBA, Coordinator
Date:	December 4, 2020
Proposal Title:	"The Prediction of Behavioral Health Outcomes in Adults with Sickle Cell Disease using the Chronic Disease Outcomes Triad Model."
	Approval # IRB-20-0123-AM01
	Reference # 108839

The Health Sciences Institutional Review Board has approved the following modification(s):

- Changed sentence in exclusion from criteria.
- Removed funding source.

There are no additional requirements in regards to your study. However, if there are further changes in the protocol after you commence your study, then you are required to resubmit your proposal for review. As a reminder, you are still require to receive continuing review and re-approval prior to your expiration date **April 3, 2023.** For further information, you may visit the FIU IRB website at http://research.fiu.edu/irb.

HIPAA Privacy Rule: N/A

Special Conditions: N/A

For further information, you may visit the IRB website at http://research.fiu.edu/irb.

MMV/em

Appendix H

Study Results and their Association with the CDOT Model

Table Legend Fully Supported Partially Supported Not Supported	
Propositions	**Associated Data**
Younger age at diagnosis of SCD will be associated with more severe hemoglobin genotypes, more complications of SCD, and increased numbers and severity of symptoms and disease consequences across the triadic domains.	90% of participants were diagnosed between the ages of birth to 7 years old. 69% of the study's participants were diagnosed with Hb*SS*. *Sample means (SD), highest score, and mode:* Pain Severity: 17.19(6.08), 30, 18. Pain Interference: 34.91(15.82), 68, 37. Presence of physical symptoms: 22.43(4.75), 35, 28. Frequency of physical symptoms: 5.68(1.93), 11, 5. Presence of psychological symptoms: 17.37(6.20), 31, 16. Frequency of psychological symptoms: 4.79(2.36), 8, 5. Depression: 10.58(8.60), 41, 6. 42% were not working or attending school and 58% were working and/or attending school.
Demographic characteristics, such as older current age, older age at diagnosis, and female gender will influence the behavioral health outcomes of persons with SCD.	Participants who were diagnosed at an older age were more likely to perceive prejudice. Males had more fatalistic attitudes than females. Current age had no significant correlation with fatalism, perceived prejudice, and self-efficacy.

	Depending on the age at diagnosis, fatalism and self-efficacy will more likely be predicted. Depending on the age at diagnosis and current age, perceived prejudice will more likely be predicted.
The more severe SCD sequealae will have a negative impact on persons with SCD who will experience more symptoms, and frequent symptoms will be experienced within and across domains, as well as have more negative behavioral health outcomes.	When participants experience severe pain, they more likely reported pain interfering with their daily lives, the presence of physical symptoms, such as weakness and pain, and high frequency of these physical symptoms. Participants with presence of psychological symptoms, frequency of psychological symptoms, and depression had effects on the participants but not significantly. When participants had increased severe pain, they will more likely report frequency of feeling sad and feeling tense or nervous. When participants reported pain interfering with their mood and ability to sleep, they more likely experienced feeling sad and had problems coping. Participants who reported weakness, pain, and yellowing of skin or eyes, also experienced the presence and severity of feeling worried or concerned, had problems sleeping, and were depressed. The more participants reported weakness, pain, and yellowing of skin or eyes, the less likely they were working and/or attending school.

	Participants who experience high pain severity, reported pain interfering with their daily lives, and the presence of weakness, pain, and swelling of hands or feet were less likely to have self-efficacy. Participants with pain, swelling of hands or feet, problems sleeping were more likely to have fatalistic attitudes and perceived prejudice. Participants who were depressed also had fatalistic attitudes and perceived prejudice. Participants with pain, number and severity of physical symptoms, depression, number and severity of psychological symptoms, and not working and/or attending school had fatalistic attitudes, although less than the study's average. Participants with pain, number and severity of physical symptoms, depression, number and severity of psychological symptoms, and not working and/or attending school, had perceived prejudice, although less than the study's average. Participants with pain, number and severity of physical symptoms, depression, and number and severity of psychological symptoms had either negative or positive self-efficacy.
The more severe and frequent triadic sequelae are, the greater will be the number of hosptializations and emergency room visits and diagnosed complications.	When participants reported severe pain, pain interfering with their daily activities, the presence of pain, yellowing of skin or eyes, and weakness, and high frequency of feeling sad, feeling tense or nervous, and problems coping, the more likely they were diagnosed

	with other SCD complications such as stroke, AVN, or eye problems. Participants with a diagnosis of other SCD complications such as stroke, painful and swollen joints, and AVN, the less likely they were working and/or attending school. Participants visited the ER with pain episodes more often when their pain was severe, increased frequency of weakness, yellowing of skin or eyes, and painful and swollen hands or feet, and increased frequency of feeling sad, feeling tense or nervous, and problems coping. Participants were admitted to the hospital for pain episodes more often when their pain was severe, the pain interfered with their day-to-day living, the presence and frequency of weakness, pain, and shortness of breath, and the presence of feeling sad, feeling worried or concerned, and problems coping. Participants with pain, number and severity of physical symptoms, depression, number and severity of psychological symptoms, and not working and/or attending school more likely had other diagnosed SCD complications such as stroke, eye problems, and AVN and experienced pain that led to ER visits. Participants with pain, number and severity of physical symptoms, depression, number and severity of psychological symptoms, and not working and/or attending school had low hospital admissions.

Appendix G

Vita

VITA

LISA GAY FRYAR

Born, Miami, Florida

1985 Jackson Memorial Hospital School of Nursing, Miami, FL

Diploma in Nursing

1997 Barry University, Miami Shores, FL

Bachelors of Science in Nursing

2000 Barry University, Miami, FL

Masters of Science Nursing (Education)

In progress Florida International University, Miami, FL

Doctorate of Philosophy Nursing, candidate

PUBLICATIONS AND PRESENTATIONS

Woodson, L.F. (2014). Living with Sickle Cell Disease: The struggle to survive. *ABNF Journal 25*(3). 86-88

2012-Nursing Education Department-Rehabilitation in Oman-Presented with Beatriz Valdes and Beverly Fray

2016-Sigma Theta Tau International (STTI)-The Health Effect with C.Framil, Tyra Tate, Scherayn Phillips-Garcia, and Dr. O. L. Strickland

2017-Sigma Theta Tau International (STTI) - Health seeking behavior of patients with PCMIs.

2019- Sigma Theta Tau International (STTI)- Chronic Disease Outcomes Triad Model