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The Lived Experience of Adults with Sickle Cell Disease During Vaso-Occlusive Crises

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THE LIVED EXPERIENCE OF ADULTS WITH SICKLE CELL DISEASE DURING VASO-OCCLUSIVE CRISES

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by

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Abstract

Background: Many sickle cell disease (SCD) patients continue to go through multiple vaso-occlusive crises (VOC) each year. The pain experienced during VOC is either undertreated or inappropriately managed because of the low attention paid by healthcare providers to SCD patients during their sickle crises. Exploring the lived experiences of adults with SCD during VOC will help the frontline healthcare providers to provide better care and develop more culturally sensitive practices that will minimize the physical and psychological suffering associated with VOC.

Purpose: This study explored the lived experience of adults with sickle cell disease, living in South Florida, during vaso-occlusive crises.

Philosophical Underpinning: A heuristic qualitative approach was guided by the interpretivist paradigm to gain an understanding of the lived experience of adults with sickle cell disease.

Methods: The sample was adults with SCD who have experienced vaso-occlusive crises in South Florida. Purposive and snowball sampling was be used. Data analysis was guided by Moustakas' (1994) transcendental phenomenology.

Results: From 20 sickle cell individuals who have experienced at least a VOC, four themes emerged *Excruciating Pain, Depressing, Feeling Helpless,* and *Dreading Healthcare Workers' Attitude.*

Conclusions: This study revealed that during vaso-occlusive crises, sickle cell individuals go through physical and psychological sufferings. The participants revealed

that healthcare workers fail to acknowledge their sufferings during VOC, and they fail to treat them effectively.

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DEDICATION

I dedicate this dissertation first to God almighty, the lifter of my head. Second, to my husband and children, I dedicate this to you, we did it!!! My husband of 26 years, I want thank you for being with me along for this journey. You told me about your multiple dreams since my FAU undergraduate years; you told me that in your dreams, I achieved my master's and doctorate degrees. You have always encouraged me to pursue higher education. You have seen my exciting moments as well as the tearful, pull my hair out, want to give up moments. In your eyes, I am a super woman and I can do a lot of things, and that has always kept me going. Your belief that I am destined for greatness which has been a source of good pressure. Every accomplishment, big or small, has always been our accomplishment because the genuine excitement you express makes it feel like you accomplished what I did with me. Sincerely, I thank you for being you. I dedicate this dissertation to God and you.

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CHAPTER ONE

Sickle cell disease (SCD) predominantly affects people of African descent living in all corners of the planet from sub-Saharan Africa, to the Middle East, India, Latin America, and Mediterranean countries (Desai et al., 2017). The disease's prevalence is highest in West Africa, where it is found in 25% of the population (Saganuwan, 2016). The high proportion of SCD in Africa can be attributed to the high distribution and carriage of the sickle cell trait (SCT) in the continent (Lee & Mark, 2014). In Africa, it is projected that more than 300 000 children are born each year with SCD, about two-thirds of them in Nigeria and the Democratic Republic of Congo (Mburu & Odame, 2019). Sickle cell disease affects about 100,000 Americans, 250 million people globally, and it is associated with major morbidity and mortality (National Heart, Lung, and Blood Institute, 2014). Most of the people with SCD in the United States are African Americans or Blacks; about 1 in 13 African American babies is born with SCT, and about 1 in every 365 black children is born with SCD (National Institute of Health, 2018).

Florida has the highest population of people with SCD in the United States with an estimate of 8,374 to 14,236 African Americans living with SCD in the state (Hassell, 2010). Despite the high number of individuals with SCD in Florida, there are only few studies on the population of people with SCD in state. Likewise, there has been no study found on adults with SCD patients during vaso-occlusive crises in South Florida. Further, research in sickle cell disease is still more focused on children than adults. Despite reductions in morbidity and mortality associated with early screening, sickle cell disease poses a substantial clinical, economic and quality of life burden to the individuals and families (Gesteira, Bousso, Misko, Ichikawa, & Oliveira, 2016). Therefore, a

phenomenological study on SCD during VOC is needed to address the adults population with sickle cell disease during pain crises.

Sickle cell disease is a group of genetic disorders that results from cells that contain an abnormal form of hemoglobin called hemoglobin S (HgbS) (Banks & Shikle, 2018). It is characterized by chronic anemia, infections, and sickling crisis. One distinctive feature of SCD is its clinical variability. Some patients develop a more severe form of SCD and suffer from intense complications and repeated hospitalizations. Other individuals exhibit more benign symptoms or are sometimes asymptomatic (Faremi, Olatubi, & Lawal, 2018). Sickle cell disease is the most prevalent and harmful hemoglobinopathy.

Sickle cell disease leads to hemolytic anemia, and patients may show signs and symptoms of hemolytic anemia, weakness, fatigue, dyspnea, including tachycardia, and long-term end-organ damage (Banks & Shikle, 2018). Patients with SCD find it more difficult to get proper care especially when they appear at the emergency department (ED) in agonizing sickle cell-related pain. There is no objective way to confirm the pain of individuals with SCD, therefore, some providers may decline to provide pain medication, which can result in patients experiencing unnecessary anguish, suffering, and a mistrust of the healthcare system. O'Connor et al. (2014) states, "many providers report feeling as though [SCD] patients exaggerate discomfort, fail to comply with medical advice, abuse drugs, or are manipulative" (p. 676). Management of SCD is complex and not well understood by most frontline providers (Darbari, 2018). Treatments for SCD are intended for support and crises prevention. A qualitative, phenomenological study is needed in order to gain an understanding of the experience of adults with SCD during

vaso-occlusive crises. Specifically, this transcendental phenomenological study is aimed at providing a description of adults with SCD from the words of sickle cell patients.

PROBLEM AND DOMAIN OF THE INQUIRY

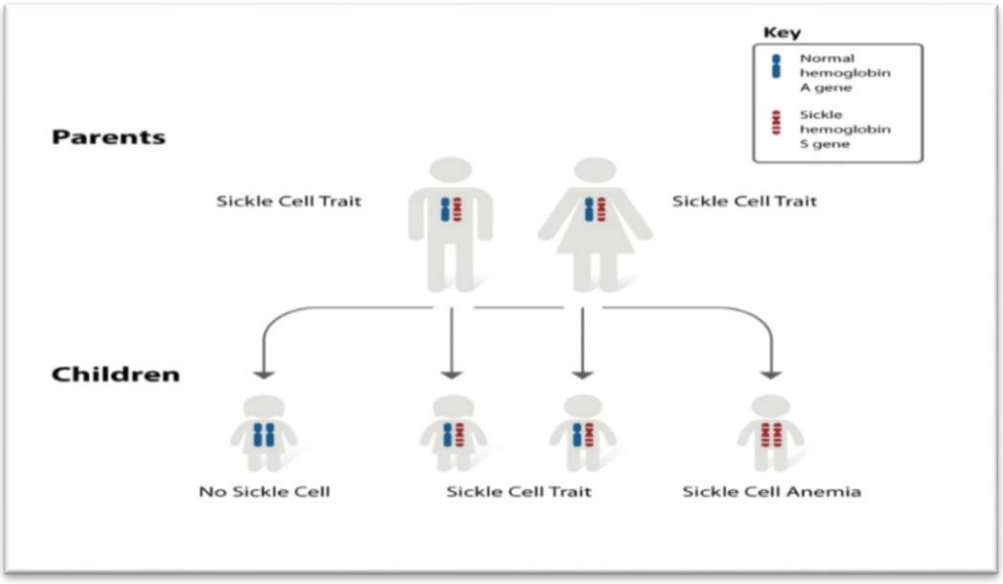
Background of the Study

Sickle cell disease, a hereditary disorder of hemoglobin, comprises a group of genetically determined conditions. Sickle cell disease is the most common of the inherited hemoglobin disorders in the world, affecting millions of people globally (World Health Organization [WHO], 2015). The chronic characteristic nature of the disease, the difficulties of prejudice, and complications that necessitate the practice of daily care can pose problems of social adaptation for people affected by this disease. Sickle cell disease management requires the daily follow-up of self-care practices, which include regular hydration, consistent medications use, frequent visits to health services, monitoring of complications, and taking care of problems related to emotional and psychosocial aspects as soon as possible (Pereira et al., 2018). The suffering, anguish, and pain associated with SCD can lead to decreased coping ability, difficulty becoming productive citizens, and a reduction in the quality of life (QOL) among those affected (Felix de Freitas et al., 2018).

People who have SCT are carriers of the sickle cell gene. When two SCT carriers make a decision to have children together, each child has a 25% chance of developing SCD. When only one parent passes the hemoglobin S gene to the child, and a normal hemoglobin gene, hemoglobin A is inherited from the other parent, the child will have SCT (National Heart, Lung, and Blood Institute, 2014). People with SCT are generally healthy. Sickle cell trait carriers have a small number of sickle cells, but enough normal

hemoglobin, hemoglobin A to avoid symptoms and complications. However, sickle cell carriers have the sickle gene that can be transmitted to their children (Harrison, Walcott, & Warner, 2016). People with SCT do not have the problems associated with SCD or experience the many complications associated with the disease. People with SCD can confirm that they have family members with SCT (NIH, 2018).

Other forms of hemoglobinopathies include hemoglobin AS, hemoglobin SC, hemoglobin D, hemoglobin E, hemoglobin O as well as beta-thalassemia. These are relatively rare forms of sickle hemoglobinopathy disease. People who have these forms of sickle hemoglobinopathy inherit one gene of abnormal hemoglobin 'S' and one gene from another type of hemoglobin such as 'D', 'E', or 'O'. Frequently, the signs and complications of these subtypical forms of hemoglobinopathies are similar in severity to those of individuals with the hemoglobin SS. People with beta-thalassemia inherit an abnormal gene of hemoglobin 'S' from one parent and a beta-thalassemia gene from the other parent. This subtype of sickle hemoglobinopathy produces signs of moderate anemia and many of the same complications associated with hemoglobin 'SS', but in a milder form. The most detrimental is hemoglobin SS; sickle cell disease (National Heart, Lung, and Blood Institute, 2018).



Inheritance Pattern for Sickle Cell Disease

Figure 1 adapted from National Heart, Lung, and Blood Institute

Sickle cell genetic disease results from the most common mutation, which is the substitution of valine for glutamic acid in the b-chain hemoglobin molecule structure. The mutation consequently results in red blood cells (RBCs) forming characteristic sickle shapes, leading to HgbS formation, at low oxygenation levels (Banks & Shikle, 2018). Sickling of red blood cells can lead to chronic blood vessel occlusion, ischemia, and immune dysfunction. Moreover, the abnormal RBCs are removed from circulation prematurely, resulting in hemolytic anemia. The diagnosis of SCD can be determined by using techniques such as electrophoresis. The clinical features and symptoms of SCD can be divided into vaso-occlusive (VOC), infectious, and anemic symptoms.

Keane and Defoe (2016) discuss that caring for a person with a chronic medical condition like SCD can be stressful for family, and is likely to have an impact on family

dynamics. If the patient is a child, the stress is most likely to be more on the parents. In developing countries, services to support parents of children with SCD are limited, and consequently, parents may feel alone and isolated (Adis Medical Writers, 2014). Following diagnosis, it is important to have adequate support and appropriate information for the families of sickle cell patients, especially the parents. It is difficult for some parents to maintain regular employment, especially mothers, while others may take what their employment can give them, so they can manage around the needs of their children (Keane & Defoe, 2016). Children with SCD may have excessive absenteeism from school because of the pain of VOC. These children may also need regular medical examinations and sick days due to pain crises of VOC. Some parents have concerns about the immediate health of their children with SCD and how to avoid painful crises. Many mothers and fathers fear the possibility that their children with SCD will have a shortened life expectancy, as well as the probability of lacking opportunities that other children enjoy and thus being unable to lead an independent life. Parents sometimes have the burden of guilt that they have passed the disease to their children (Keane & Defoe, 2016).

The treatment of SCD that black people receive is affected by the health disparity that continues to exist among people of diverse ethnic and socio-economic backgrounds in the United States. Due to the historical stigmatization of people of African descent in particular, black people face significant health inequality (Jenerette, Brewer, & Ataga, 2014). People of African descent have been marginalized and disallowed to participate in mainstream society since being sold into the trans-Atlantic slave trade. The legacy of this level of ill treatment still makes it difficult for them to access adequate healthcare (Cunningham et al., 2017). Families of children with SCD usually report that they have

difficulty: accessing care; reaching providers by phone; getting an appointment promptly; not being delayed during visits; and problems paying for prescriptions (Crosby et al., 2015). Some of the reasons for improper care during crises can be related to stigmatization and health disparity. Individuals with SCD are more likely to experience discrimination in the health care system particularly around treatment for pain; it is believed that the health disparity is related to ethnic and racial issues (Gesteira, Bousso, Misko, Ichikawa, & Oliveira, 2016).

Many adults with SCD might have health care insurance, usually Medicaid or Medicare, or both (Hulihan, Hassell, Raphael, Smith-Whitley & Thorpe, 2017). Sometimes, gaps in coverage might prevent sickle cell patients from accessing care. High deductibles might prevent use of services or insurance plans might not cover some essential services (Hulihan, Hassell, Raphael, Smith-Whitley & Thorpe, 2017). Irregular health insurance coverage can arise if there is loss of a job that provided the insurance because of the disability from SCD or if the patient gains a job that does not provide health insurance. Inability to access proficient providers and optimal care can additionally complicate living with sickle cell disease and limit treatment options (Hulihan, Hassell, Raphael, Smith-Whitley & Thorpe, 2017).

There are very few treatment modalities for SCD and currently the treatment of symptoms seems to be the most viable option. Nevertheless, bone marrow transplantation is a promising cure for SCD with a 91% event-free survival rate after 5 years of follow-up and 93% survival rate, when there is a matching donor (Hulihan, Hassell, Raphael, Smith-Whitley & Thorpe, 2017). Unfortunately, only one in ten sickle cell patients is able to find a matching donor (Sickle Cell Association of Ontario, 2018). Only 1,000

individuals with SCD globally have received an identical sibling transplant (Hulihan, Hassell, Raphael, Smith-Whitley & Thorpe, 2017). Hydroxyurea, an antineoplastic drug, has also been used in the treatment of SCD, but the medication only suppresses the frequency of VOC (Nottage, 2016). Sickle hemoglobinopathies are a major health problem, which places an immeasurable emotional, psychological, and economic burden on families. All healthcare professionals should understand the impact of VOC as this may assist them to provide more compassionate care and help prevent associated complications of morbidity and mortality.

The need of people with SCD must be met as quickly as possible at the emergency unit, in a heartfelt, sincere, and compassionate way, by a competent healthcare team. Their need has to be met as they appear in the ER in a weakened state related to the painful episode of VOC and or the manifestation of other complications that limit their activities and impact their QOL. The most common complications that make the person with SCD seek ER services are pain, anemia, infection, and stroke, which necessitate a careful evaluation and fast intervention by the healthcare providers. Therefore, the care of the persons with SCD in the ER involves complex measures such as individualized evaluation, pain management, proper understanding of immunologic and hematologic issues, and prevention and control of infection. (Carvalho, Santos, Izidoro, Caldeira dos Santos, & Batista Santos, 2016).

While there is no FDA approved readily accessible cure for SCD (except for bone marrow transplantation where there is a matching donor), early finding through counseling programs, genetic screening, and educational programs can help prevent and/or manage the crises. Routine screening during prenatal period and at birth can

identify the disease as early as possible (WHO, 2015). In order to meet the serious challenges associated with caring for patients with SCD, it is often necessary to forge partnerships between healthcare providers, the scientific community, governments, and nongovernmental organizations (Adis Medical Writers, 2014).

Pathophysiology of Vaso-Occlusive Crises

Vaso-occlusive crises result from adhesion of low-density sickled red blood cells and reticulocytes in immediate postcapillary venules leading to trapping of the older, denser, and distorted sickled red cells, resulting in reduced blood flow. It was found that the random precapillary obstruction by a small number of thick sickled red cells also contributes to VOC (Manwani & Frenette, 2013). The damaged sickled red cells activated endothelial cells, and intravascular hemolysis lead to inflammation, which results in coagulation. This adhesive interaction of damaged red cells and leukocytes to the endothelium plays an important role in the initiation of VOC.

Most people who suffer from SCD experience frequent VOC and pain. On average, 55% of patients with SCD experience a VOC at least once or twice a year while 5% may have a VOC three to ten times per year, and 1% have greater than ten VOC per year (Carroll, 2015). Factors such as hereditary and acquired factors contribute to this clinical variability; socioeconomic status emerges as one of the leading factors affecting clinical manifestations of VOC (Adenike. Matthew, & Lawal, 2018). Many healthcare providers tend to underestimate the suffering and agony that persons with SCD experience during the time of VOC (Franceschi et al, 2016).

Vaso-occlusion can result in osteonecrosis, acute chest syndrome, organ failure, skin ulcers, and cerebrovascular accidents. The sickling of red blood cells results in their shortened lifespan in the circulation and characteristic anemia (Gardner, Boll, Bhosale, & Jaffe, 2016). For acute non–life-threatening VOC episodes, immediate therapeutic treatment comprises of hydration, applying heat, anti-inflammatory agents, antibiotics, and opioids, which are useful in improving oxygen delivery to tissues and controlling pain. Vaso-occlusive crises cause abdominal pain, which is one of the principal reasons SCD patients seek medical attention. Gardner, Boll, Bhosale, and Jaffe (2016) assert that these abdominal pain episodes are often attributed to microvascular occlusion having led to infarcts in the mesentery and abdominal viscera, which can affect the spleen, liver, biliary system, kidneys, and gastrointestinal tract.

Sickle cell disease's painful episodes lead to a substantial number of hospital admissions and readmissions each year. Vaso-occlusive crises lead to 197,000 ED visits and 356 million dollars spent on pain management every year, and many patients with SCD visit the ED repeatedly for pain management because VOC are typically recurrent (Lentz & Kautz, 2017). These frequent admissions account for the majority of health care expenses associated with the disorder. The more frequently the visits to hospitals, the more likely the fatality and mortality will be (Pizzo et al., 2014). Activities of daily living (ADL) are adversely impacted during VOC, as well as the ability to stay asleep.

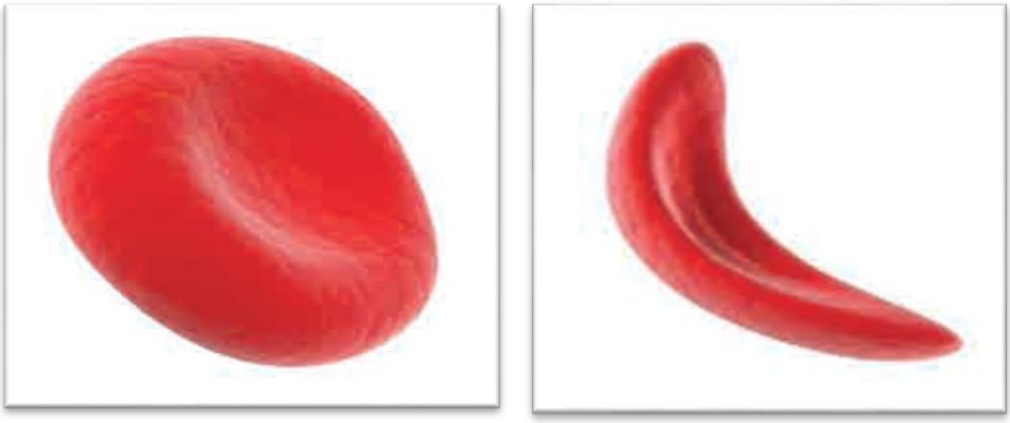


Figure 2: Normal (A) and sickled (B) red blood cell shapes. Adapted from Community Practitioner, June 2016.

Complications of Sickle Cell Disease

Patients with SCD suffer from the following complications: unpredicted pain episodes of VOC, chronic anemia, acute chest syndrome, pulmonary hypertension, kidney disease, gallstones, strokes, and long-term end-organ damage (Gardner, Boll, Bhosale, & Jaffe, 2016; Hamm et al., 2016; Hulihan, Hassell, Raphael, Smith-Whitley, Thorpe, 2017). These SCD-related complications result in significant impairment in QOL. Likewise, SCD has economic complications such as enormous frequent hospitalization, lack of education, unemployment, poverty, neglect, and disability (Damille Silva et al., 2014). Sickle cell disease as a chronic and limiting disease can interfere with patients' ability to perform their daily activities. The inability to perform daily activities may lead to permanent or temporary disability, as well as irreparable harm to the functioning of several organs, causing work impairment and leading to a decrease in QOL. Sickle hemoglobinopathies are associated with lower scores on QOL (Felix de Freitas, 2018).

Sickle cell disease is harmful to the whole body. According to a recent report by the National Institutes of Health (NIH) (2018), the cardiopulmonary system of a sickle cell patient is overworked and compromised due to its failure to adequately supply the body with healthy red blood cells necessary to maintain sufficient levels of oxygen throughout the body. Over a lifetime, SCD can harm a patient's spleen, eyes, brain, liver, lungs, heart, penis, kidneys, joints, bones, or skin (National Heart, Lung, and Blood Institute, 2018). Vaso-occlusive crises episodes can cause problems and negative experiences for people living with SCD. The report from the NIH found that people living with SCD have reduced physical ability, from minor discomfort to complete inability to perform daily activities. Vaso-occlusive crises may cause suffering and generate feelings of negative self-image. It may cause anxiety or a vague feeling of apprehension, low self-esteem, social rejection, and social isolation. It may also cause depression or overwhelming sadness, feelings of hopelessness, apathy, and substandard performance in school or work (Adzika, Glozah, Ayim-Aboagye, & Ahorlu, 2017).

Leg ulcers have been found to be one of the chronic complications of SCD that tend to be hard to treat. Their slow healing process can severely interrupt quality of life. The pathogenesis may include numerous factors, such as mechanical obstruction by dense sickled red cells from hemolysis, infections, venous insufficiency, anemia and its resulting decreased oxygen-carrying capacity (Rivolo, 2018).

Sickle cell disease can negatively impact sexuality in men. Priapism is a complication that affects the sexuality of males with SCD. It has been estimated that 40% of adult men with SCD experience at least one incident of priapism during their lifetime (Costa et al., 2018). Priapism is a disorder in which a man gets a persistent erection, not

accompanied by sexual stimulation or desire, which usually lasts for more than six hours, and characteristically involves only the cavernous body of the penis. Priapism can lead to sexual impotence which negatively impacts sexuality, self-care, and QOL (Costa et al., 2018). In patients with SCD, priapism is related to low hemoglobin levels and changes in hemolytic activity. Priapism is a urological emergency, and its inadequate management can lead to erectile and sexual dysfunctions (Costa et al., 2018). This condition requires knowledge from the health care providers to give proper care and promote self-care.

End-stage renal disease (ESRD) affects individuals with SCD. The incidence of ESRD in SCD adult population has been estimated to be between 4% and 12%, and 20%–40% of patients with SCD related chronic kidney disease (CKD) progress to ESRD (Boyle, Jacobs, Sayani, & Hoffman, 2016). Chronic kidney disease results from the effects of recurrent episodes of red blood cell sickling in SCD patients. Glomerular injury defined as an estimated glomerular filtration rate (eGFR) of <90 manifests in adults with SCD, with up to 50% of them showing signs of hyperfiltration, and 21%–27% having CKD. The injury affects the glomeruli, leading to global or segmental glomerulosclerosis, glomerular hypertrophy, and duplication of the glomerular basement (Mammen, Matsell, & Bissonnette, 2017). Sickle cell disease patients develop significant long-term, progressive, and permanent kidney injury.

Acute chest syndrome (ACS) is a recurrent complication of SCD in patients hospitalized with vaso-occlusive crises. Acute chest syndrome is a type of critical lung injury in SCD patients. Acute chest syndrome is the second most common cause for hospitalization after VOC in patients with SCD (Jain, Bakshi, & Krishnamurti, 2017). The development of ACS represents a cycle of lung infarction, swelling, and atelectasis

leading to, hypoxemia, ventilation-perfusion mismatch, and acute increases in right ventricular and pulmonary artery pressures. It is associated with a high risk of sickle cell-related morbidity and mortality in children, including prolonged hospitalization (Bundy et al., 2017). More than half of all children with hemoglobin SS experience at least one incident of ACS in the first ten years of life. Repeated episodes of ACS may cause the beginning of debilitating chronic lung disease. The major identifiable etiologies for the development of ACS are pulmonary infarction, pulmonary infections, and pulmonary fat embolism from necrotic bone marrow (Nansseu et al., 2015).

Pulmonary infarction is a significant contributor to the development of ACS and was determined to be the cause in 16% of ACS episodes in National Acute Chest Syndrome Study (NACSS). Pulmonary infarction occurs as a result of increased adhesion of sickled red blood cells to the endothelium resulting in vaso-occlusion, which leads to worsening of hypoxemia and ventilation-perfusion discrepancy. Pulmonary infections can cause an ACS episode in a vulnerable SCD patient by inducing an extreme inflammatory lung injury response in the presence of an injured lung microvasculature (Thomas & Simmons, 2017).

Pulmonary fat embolism, with or without pulmonary infection, can precipitate ACS in both children and adults. During acute VOC pain episodes, necrosed and infarcted bone marrow (especially of the pelvis and femur) release fat droplets in the bloodstream, which embolize to the lungs. Fat emboli in the lung vasculature are metabolized to free fatty acids, including secretory phospholipase, which mediates endothelial injury and alveolar inflammation. There is a high occurrence of pulmonary

embolism in SCD patients associated with thrombosis in the distal venous circulation (Nansseu et al., 2015).

Acute chest syndrome can present like asthma exacerbation, bacterial pneumonia or bronchiolitis at presentation. It can likewise progress quickly from mild hypoxemia to acute respiratory failure within 24 hours of onset of respiratory symptoms. This type of rapidly progressive ACS is more commonly seen in adults with SCD and is frequently associated with multiorgan failure. Recurrent episodes of ACS in childhood may lead to fibrosis, chronic hypoxemia, obstructive and restrictive abnormalities, and early mortality (Thomas & Simmons, 2017). .

Other complications of SCD include hypertension, stroke, epidural hematoma, transient ischemic attacks, hepatic sequestration, and cholelithiasis (Barriteau & McNaull, 2018). People with SCD can also develop heart murmur, cardiomegaly, and congestive heart failure. Peripheral neuropathy, pulmonary hypertension, chronic pulmonary disease, delayed growth and puberty, premature labor and delivery, and permanent disability can also be potential complications. Repeated vaso-occlusive crisis is the most frequent complication of SCD (Gardner, Boll, Bhosale, & Jaffe, 2016).

Treatment of Sickle Cell Pain

Matthie and Jenerette (2015) declare that pain is a subjective phenomenon that shows in various ways, and that nurses are taught that pain is whatever the person is experiencing it says it is, existing whenever he or she says it does. Sickle cell patients usually describe their pain as sharp, stabbing, dull or throbbing, intense, and in most cases is located in the chest, arms, abdomen, back, knees, and legs. There are no objective findings to support the presence of a pain crisis and the pain reports. Healthcare

providers must depend on the patient's report to efficiently assess and the pain of SCD because of the individual nature having no objective findings (Matthie & Jenerette, 2015).

The undertreatment of pain in the ED is a substantial medical and ethical issue. Under-treated acute pain contributes to poorer physical and mental health outcomes, and increases the risk of chronic pain (Carter, Sendziuk, Eliott, Braunack-Mayer, 2016). Access to proper pain management is gradually conceptualized and promoted as a human right. Per International Association for the Study of Pain (IASP), all people have the right to have their pain recognized and be informed about how their pain can be managed, and to have access without discrimination (IASP, 2015). Vaso-occlusive episodes necessitate opioid treatment. In spite of the indication to support speedy pain management within 30 minutes, with reassessments done every 15-20 minutes after a pain medication is given until the patient reports a bearable pain level, care for SCD patients in VOC does not reliably meet this benchmark (Kim, Brathwaite, & Kim, Ook, 2017).

Healthcare providers are very aware of substance abuse and drug-seeking behaviors among some patients. Hence, patients who frequently visit the ED for VOC pain management may undergo special scrutiny to determine whether their pain is actually present, or if their pain is from a physical or psychological illness. It has been found that some frontline healthcare providers have a negative attitude toward patients with SCD, especially during a VOC visit (Kim, Brathwaite & Kim, Ook, 2017). The negative attitude of providers has caused slower pain treatment, and in some cases, a diversion from national guidelines for proper care for patients with SCD. Patients with VOC have conveyed significant negative hospital experiences, characterized by delays in

pain control, mistrust, lack of autonomy over their treatment, lack of psychosocial support, stigmatization, poor monitoring, and neglect (Matthie & Jenerette, 2015). Patients have reported that they feel less than human when trying to obtain medical care needed during a vaso-occlusive episode (Lentz & Kautz, 2017).

There is mistrust from both patients and the health care providers. This mistrust creates barriers between SCD patients and their providers, and this affects how they each react when confronted with a VOC (Wakefield, Zempsky, Puhl, Litt, 2018). For healthcare providers, opinions of drug-seeking and addictive behaviors affect their assessment of these patients, negatively impacting the relationship. These negative attitudes from healthcare providers and their inadequate knowledge of VOC have been recognized as serious barriers to effective pain management in SCD and have led to poor adherence to current pain management protocols (Bergma & Diamonf, 2013. Patients foreseeing and anticipating this skeptical attitude may delay seeking medical care for VOC. Failure to treat a VOC speedily and appropriately can lead to debility or death from complications (Gardner, Boll, Bhosale, & Jaffe, 2016).

Many ED physicians have become skeptical about who gets opioids in the ED and who gets a prescription to go home with. This reluctance can result in unnecessary suffering for sickle cell patients who require opioids to relieve their pain caused VOC. Sickle cell disease patients who present to the ED with acute pain crises associated with SCD experience significantly lengthier delays of the administration of the initial pain medicine compared to other patients with similar pain despite higher arrival pain score and triage acuity levels (Lazio et al., 2010). Besides, there may be other consequences apart from suffering; distrust of the medical system is also a big problem, and the patients

may be reluctant to seek care in the future during VOC because of their previous experience with their health care providers. Some physicians may overestimate the prevalence of addiction or opioid abuse or use wrong criteria to judge the severity of SCD pain reports. Besides inappropriate fear of addiction or opioid abuse, some doctors suspect that underlying racism partially explains undermedication of SCD patients (Smith, 2014). This is supported by disparities in prescribing opioids in general to blacks versus whites though the prevalence of prescription abuse in whites is significantly higher than Blacks (Berezow, 2018). When classified by race/ethnicity, the drug-related mortality rate (per 100,000) in 2016 for whites was 25.3, blacks 17.1, and Hispanics 9.5 which shows that whites were roughly 50% and 167% likelier to die from drug overdoses than Blacks and Hispanics, respectively (Berezow, 2018).

A treatment protocol was developed for SCD pain in adults who appear in the emergency departments that allows for patient controlled analgesia (PCA) for individuals awaiting hospitalization. Myers and Eckes (2012) asserted that the use of PCA has positive outcomes for patients in both pain management and satisfaction, especially during pain crises. Optimal treatment of vaso-occlusive pain episodes involves bolus dosing of intravenous opioids; frequent reassessments of pain; and close observation of the patient. All of the aforementioned interventions are time consuming for ED nurses and can lead to delays in the care of patients with SCD. Delays frequently occur when the patient's condition necessitates hospitalization for continued analgesia while waiting in the ED before an available hospital bed (Santos, Jones, Andemariam, Wkefield, Grady & 2016). Although the American Pain Society guidelines recommend PCA for managing pain during VOC, the initiation of PCA is often delayed. Thus, patients usually do not

receive PCA until after being transferred to the hospital floor extending their suffering from VOC associated pain (Santos, Jones, Andemariam, Wkefield & Grady, 2016).

Quality of Life in Sickle Cell Disease

Quality of life includes many aspects which comprises areas related to mental, emotional, physical, and social functioning. The complications of SCD: hemolysis, and vaso-occlusive crises with repeated painful episodes, stroke, splenic sequestration, cholelithiasis, and acute chest syndrome, and that can lead to life-long disabilities and death can lead to significant impairment in QOL (Cevher, Unal, Kucukkavruk, Tasdelen, & Tunctan, 2018). Problems with the duration of treatment, combination of physical, social, and psychological issues, in addition to comorbid diseases and associated drug use seem to be the common causes of poor compliance to treatment which leads to poor quality of life.

There is considerable impairment of quality of life in adults with SCD who are mostly in severe pain, are receiving blood transfusions, and hospitalized. The poor quality of life in SCD patients might be related to general physical limitations and lower daily functioning abilities. Besides, the factors that affect and threaten the survival of patients with SCD have a negative effect on their quality of life (Adzika, Glozah, Ayim-Aboagye & Ahorlu, 2017). Factors such as age, social relations (marital status), and levels of education (income) should be considered seriously in the management of SCD to facilitate healing and improve quality of life. Vaso-occlusive episodes, the clinical hallmark of SCD, are a medical emergency, debilitating, and cause poor quality of life.

Disability form the disease and inability to perform activities of daily living can worsen the QOL of individuals affected by SCD. Pain episodes from VOC, physical function mobility, fatigue, depression, and anxiety, affect QOL in sickle cell disease. Individuals with SCD have more recurrent hospitalizations and more frequent emergency room visits compared to other people with other chronic disease conditions (Badawy, Thompson, Barrera, Cai, 2018). Vaso-occlusive pain is the most frequent and stressful SCD complication and is related to poor QOL. Adults and youth with SCD report lesser QOL in various areas of health compared with other people with other chronic conditions. Pain of SCD becomes more chronic and persistent as children get older (Ludwig, Sil, Khowaja, Cohen, & Dampier, 2018).

Sickle cell patients are usually dissatisfied with their quality of life. Poor QOL affects life expectancy of persons with SCD; life expectancy in individuals with SCD is reduced significantly (Adzika, Glozah, Ayim-Aboagye & Ahorlu, 2017). Children with SCD may not perform well in school because of excessive absenteeism related to complications from SCD. Excessive absenteeism may eventually lead to school dropout. School dropout impacts the socio-economic circumstances of people with SCD, and this may lead to a negative effect on the capability to manage SCD. Socio-economic distress can predict QOL; living in a troubled neighborhood predicts reduced health-related QOL in SCD individual (Adzika, Glozah, Ayim-Aboagye & Ahorlu, 2017). The chronicity of sickle cell disease, the accompanying frequent hospital visits, appearance, and poorer physical state of the individual affected may also contribute substantially to impairment in the quality of life and wellbeing of their family members (Wonkam, Mba, Mbanya, Ngogang, Ramesar & Angwafo III (2014).

Statement of the Problem

People with SCD during VOC should receive prompt and adequate care, especially for pain management. However, many frontline providers fail to provide speedy and adequate care for this group of people during crises due to disbelief or skepticism in pain reports and the possibility of addiction and or abuse (Carroll, 2016). A chronic illness such as SCD can make people affected feel restricted in their day-to-day activities and can result in mortality if proper care is not given during crises (Adeyemo, Ojewumi, Diaku-Akinwumi, Ayinde, & Akanmu, 2015). Health care providers should be aware of the physical and psychologic suffering experienced by the individuals affected by sickle cell during VOC. According to the American Nurses Association (ANA) code of ethics, nurses have a responsibility to protect and advocate for the health and safety of their patients (ANA, 2015). By understanding the experience of SCD sufferers during VOC, nursing will be able to provide adequate care in relation to pain management.

Purpose of the Study

The purpose of this qualitative, heuristic phenomenological study was to explore the lived experience of adults with sickle cell disease living in South Florida, during vaso-occlusive crises. This research aimed to give adults with sickle cell disease a voice to express their individual experience during vaso-occlusive crises, to provide an inductive description of the phenomenon, and to gain the understanding of the essence of the experience of vaso-occlusive crises.

Research Question

The research question of this study was: What is the lived experience of adults with sickle cell disease living in South Florida, during vaso-occlusive crises?

Philosophical Underpinning

Once an issue to be examined has been identified, the researcher must determine how to investigate the issue. The researcher begins by finding the paradigm in which to position the study. A paradigm is a worldview that seeks to develop an understanding of the human experience using a set of interpretive practices that make the world visible (Kuhn, 2012). Data may be collected through a variety of quantitative and qualitative research methods that may balance each other and move knowledge closer to the truth. Though SCD can be examined using either quantitative or qualitative method, the approach that was suitable to examine the lived experience of adults with sickle cell disease during VOC was the qualitative approach using interpretivism and constructivism paradigm.

Post-Positivism

Post-positivism offers an alternative to the traditions and foundations of positivism for conducting an inquiry. Post-positivism arose to address the inadequacies perceived by researchers in the application of the traditional methods of positivism. For the post-positivist, reality is not rigid; It is a creation of those individuals involved in the research. Post-positivism believes in reality and a want to understand it, sees objectivity as a goal, but also identifies the impossibility of total objectivity (Polit & Beck, 2017). Post-positivists realize the impossibility of total objectivity. They believe that reality does not exist in emptiness, its composition is influenced by its context, and many constructions of reality are therefore possible. Culture, gender, and cultural beliefs are important factors that affect reality. Popper (1902-1994) argues that certainty in knowledge was an illusion, because given the universal of scientific claims of scientific

theories, we can never demonstrate them on the basis of our particular experiences. The ontology is critical realism, as claims about reality must be subjected to critical investigation to facilitate the uneasiness of reality. While post-positivism continues to consider the metaphysical as being beyond the scope of science, they also believe that it is impossible for humans to truly perceive it with their imperfect sensory and mental capacity.

Interpretivism

Interpretivism is often linked to the belief of Max Weber who suggested that people are concerned with understanding in contrast to the explicative approach of explaining (Crotty, 2013). Interpretivism looks for culturally derived and holistically situated interpretations of the social life-world. Interpretivism aims for understanding, not cause and effect. Interpretivism pursues to understand the unique development of phenomena as experienced by the individual, not to be able to generalize the phenomena and apply them to everyone. Interpretivism relies mainly on inductive reasoning.

Interpretivism depends greatly on naturalistic or qualitative methods such as interviewing and observation which warrant an adequate dialogue or discussion between the researchers and the people they interact with in order to construct a meaningful reality. Interpretivism emphasizes on language, symbolism, and meaning, rather than investigating a phenomenon with surveys and measurement instruments that decrease human activity to statistically quantifiable pieces. Interpretivism is guided by constructivism and constructionism (Crotty, 2013).

Constructivism

Constructivism assumes that individuals create their own reality, their own meaning, through their understanding of personal actions and interactions with objects and other individuals in the world. Constructivism is a belief that there is no objective truth waiting to be discovered (Crotty, 2013). Truth and meaning only come into being through the engagement of the realities. From a constructivist viewpoint, different people may construct meaning in diverse ways, even concerning the same phenomenon. These meanings are constructed by people as they engage with the world they are interpreting (Crotty, 2013).

In constructivism, the relationship of the researcher to the participant is inseparably intertwined in knowledge production. Subjectivism, the epistemological underpinning for constructivism, integrates human interest into a study, and it is concerned with the understanding of the issue being studied. “Once we understand the interior subjective world of the individual or culture, we have accomplished the first step of knowing the truth, the meaning, and the interpretation from the source” (Munhall, 2012, p. 21).

Researchers using constructivist paradigm use inductive reasoning to investigate issues. Inductive reasoning involves commencing from a series of precise cases and ending up with a general statement. The researcher collects data through interviews, observation, and then analyzes the data. Information is gathered by talking directly to the participants, collecting data face-to-face from the participants, and watching their behavior through interaction with them. The researcher works continuously between the themes and the database until a comprehensive set of themes is established. The researcher sometimes

may need to collaborate with the participants giving them a chance to shape the themes that emerge from the process (Creswell & Poth, 2018).

Constructionism

Constructionism philosophical underpinnings claim that meaning or truth is not discovered or created but constructed as people interact in a world in which they live (Crotty, 2013). Constructionism is the view that all knowledge and all meaningful reality is dependent upon human practices, being constructed in and out of the interaction between human beings and their world. Constructionism maintains that meanings are constructed by people as they involve with the world they are interpreting. Constructionism believes that people do not create meaning, they construct it. Constructionism believes that objectivity and subjectivity need to be brought together and held together inseparably. Constructionists view knowledge and truth as created, not discovered by the mind. Constructionists concern themselves with what is known about the human world of social experience, not the objective realities of the natural world (Walker, 2015).

Constructionism emphasizes on the unique experience of each person, one's individual way of making sense of the world is as valid and worthy of respect as any other individual. Social constructionism points out the influence culture has on the way in which one sees and feels things and defines how one sees the world. One depends on culture to direct our behavior and organize our experience. Culture is best realized as the source rather than the result of human behavior, and without it, one could not function (Crotty, 2013). Constructionists do not deny the presence of reality, but they uphold that its meaning is socially constructed (Walker, 2015).

Qualitative Research

When seeking to understand meaning, make sense of, or interpret a phenomenon, a qualitative approach is used. Qualitative research is designed to understand a phenomenon. Creswell and Poth (2018) defined qualitative research as a situated activity that discovers the observer in the world. They further stated, "It consists of a set of interpretive and material practices that make the world visible, and that involved an interpretive, naturalistic approach to the world" (p.7). Qualitative research is used for many reasons. It is used to explore or examine a phenomenon of interest, hear quietened voices, and variables that cannot be easily measured. Qualitative research is conducted when a complex and detailed understanding of an issue is needed. It is also used when quantitative procedures and statistical analyses do not fit the problem (Creswell & Poth, 2018). Qualitative inquiry uses different approaches that researchers can use to describe and understand individuals' subjective worlds that emerge from their experiences (Munhall, 2012). Qualitative research is used to develop theories when partial or inadequate theories exist. Qualitative research starts with assumptions and the use of interpretive and theoretical frameworks, and the exploration of the meaning of the human experience (Creswell & Poth, 2018).

The characteristics of qualitative studies are identified by Creswell and Poth (2018) as follows:

1. The researcher collects data in natural settings where the participants experience the issue under study.
2. The researcher is the main instrument in collecting the data.

3. Qualitative researchers use multiple forms of data (interviews, observation or documents).
4. Qualitative researchers use complex reasoning through inductive-deductive logic process
5. It focuses on the participants' multiple perspectives and their meanings.
6. The researcher is located within the setting of participant or sites
7. Reflexivity; the researcher is positioned in the research, conveys his background and how personal experiences informs the investigation.
8. A holistic view is embraced (complex interactions are identified).
9. An emergent design is used.

There are five main approaches in qualitative research: phenomenology, narrative research, grounded theory, ethnography, and case study (Creswell & Poth, 2018). Qualitative research starts with assumptions, identification of a paradigm, and exploration of the meaning of the human experience. These assumptions are stances that the researcher takes to provides direction for the study (Creswell & Poth, 2018). They are important to the philosophical underpinning of qualitative research in which paradigms are built (Munhall, 2013). Assumptions as underlying principles that are assumed to be true. The scientific assumptions are ontology, epistemology, axiology, rhetorical, and methodology (Elsbach & Kramer, 2015).

The ontological assumption relates to the nature of reality and its characteristics. The ontological assumption in interpretivist qualitative research is that reality is relativist and contextual. In qualitative research, it is believed that there are multiple realities. Nevertheless, post-positivist research is underpinned by a different ontological

assumption: there is only one reality, but it can only be partially known, and the researcher may not be able to fully comprehend because of lack of absolutes (Creswell & Poth, 2018). When studying people, qualitative researchers conduct studies with the intent of reporting these multiple realities (Creswell & Poth, 2018). It is believed that realities are constructed through our lived experiences and interaction.

The epistemological assumption comes from how knowledge is known. In interpretivist qualitative inquiry, knowledge is subjective as seen by participants. However, in post-positivist qualitative inquiry, knowledge is generated objectively. Epistemology assumption is built on the basis that reality is co-constructed between the researcher and the participants and are shaped by their experiences. The researcher attempts to get as close as possible to the researched; therefore subjective evidence is obtained from participants based on individual views. When the researcher stays in the field longer to get to know the participant more, they have the opportunity to know more what they know (Creswell & Poth, 2018). In heuristic phenomenology, the researchers seek descriptions of the phenomenon that reveals its basic structure, rather than interpretations that focus on the meaning of the phenomenon. The researcher uses epoche to provide the analytical distance necessary to maintain objectivity.

Axiological assumptions answer the question: What is the role of values in research? All qualitative researchers bring values to research and make their values known in a study. Intuition and biases are important, though the researcher will understand that the participants' responses take precedence over the preconceptions and biases of the researcher. Participant's subjective ideas are valuable. The researcher may use bracketing to account for researcher bias. Bracketing is setting aside preconceptions,

biases, and preconceived ideas to take a fresh viewpoint toward the phenomena under investigation. The post-positive researcher's biases are controlled and not expressed. Values are excluded from the research process.

The rhetorical assumption is the language of the research. It involves analyzing, using, and developing principles and criteria of evaluation within subjects. In qualitative studies, the language is often informal; it is often written informally, in the first person. The post-positive researcher writes from the perspective of the disinterested scientist. Typically, the report is couched in mathematical terms.

The methodological assumption in qualitative research involves the use of inductive reasoning to get ideas through ways like interviews, observations, and symbols and analysis the collected. The phenomenon of study is studied in its natural setting data instead of collecting data in a laboratory. During data analysis, the researcher follows a path of analyzing the information to develop a more detailed and thorough knowledge of the phenomenon being studied. The research method should align with the topic and address the research question (Creswell & Poth, 2018). In post-positivism research, scientific technique is used, and deductive approaches are important, such as testing of theories, specifying important variables, making comparison among groups (Creswell & Poth, 2018).

In the context of this study, the ontologic assumption would be: What creates reality during vaso-occlusive crises in adults with sickle cell during? The epistemological assumption would be: What can be counted as knowledge in the lived experience of adults with sickle cell disease during vaso-occlusive crises? The study would ask: Are there means to access knowledge of the reality that adults with sickle cell disease make of

their world during vaso-occlusive crises? These questions determine the methodological approach. The decisions made during data collection are important to the overall coherence of the study and rely on application of the method. It is important that the methodology is compatible with the topic and that it addresses the research question. There are several qualitative approaches; however, this study will use phenomenology as a guiding framework. The above assumptions will be used to guide the phenomenological philosophical underpinning.

Phenomenology

A phenomenological study describes the common meaning for several individuals of their lived experiences of a concept or phenomenon (Creswell & Poth, 2018, p. 75). Phenomenologists concentrate on describing what all participants mutually have as they experience a phenomenon. The phenomenology approach is to suspend all judgments and findings about what is factual (the natural attitude) until they are founded on more certain basis (Creswell, 2013). Interpretation and meaning of different things depend on the context in which they are perceived. The perception of the lived experience should be interpreted from the individual's unique perspective (Munhall, 2012).

Phenomenology focuses on the experiences of the individuals and the phenomena that appear in the consciousness; thus, people's perceptions and experiences of the same phenomena will vary (Matua, 2015). Consciousness is sensory awareness of and response to the environment. The unity of mind and body becomes a means of experiencing, eliminating the idea of a subjective and objective world (Munhall, 2012). Embodiment explains that through consciousness individuals are aware of being-in-the-world, and it is through the body that we gain access to this world. We are thought of as being-in-the-

world because we participate in cultural, social, and historical contexts of the world (Munhall, 2012). The natural attitude is a mode of consciousness that promotes interpreted experiences. The world as experienced and interpreted by preceding generations is handed down, and these teachings become a part of a person's natural attitude toward the world (Munhall, 2012). Reality is inseparably related to one's consciousness of it because consciousness is indivisible from and unable to exist independently of its object (Matua, 2015).

Phenomenology is often described as a philosophy and a research method. Two major types of phenomenology are Husserl's descriptive, transcendental phenomenology, and Heidegger's interpretive phenomenology. Husserl introduced phenomenology in the mid-1980s, and he had the belief in the meaning of a person's experience(s). He also held the view that realities are unpolluted phenomena from which all studies must begin (Munhall, 2012).

Heideggerian Phenomenology (Hermeneutic)

Heidegger, Sartre, and Merleau-Ponty expanded on Husserl's view. They examined the lived world of people and paid more attention to *essences* of that world. Martin Heidegger believed that cultures, history, and worldviews influenced the interpretation of meaning (Creswell & Poth, 2018). Interpretive or hermeneutical phenomenology is described as research concerned the interpretation of the meaning of the importance of one's experience through which themes are formed. Van Manen described research as oriented toward lived experience. He believed that phenomenology is not only a description but also an interpretive process in which the investigator makes an interpretation of the meaning of the lived experiences (Creswell & Poth, 2018).

Martin Heidegger was a student of Husserl and together they brought a cognizance to phenomenology. Heidegger worked with Husserl, who taught him in the processes of phenomenological intentionality and reduction. He became so efficient in his work that Husserl assumed he had found the successor he had been looking for, and he ensured Heidegger's succession to his professorship even though Heidegger disassociated himself from Husserl and his work. Heidegger then began a radical reinterpretation of Husserl's phenomenology (Van Manen, 2014). Heidegger rejected the theory of knowledge known as epistemology and adopted ontology, the science of being. Heidegger believed in being-there of human existence in a specific social world, which is constructed by a set of historical, cultural, and social meanings. Heidegger believed that interpreting meanings is influenced by cultures, history, and worldviews. Heidegger believed that human existence found itself in-a-shared-world-along-side-other, and that these others are not configured as subjects or alter egos but as the very ontological character by which human existence is revealed. Heidegger developed interpretive phenomenology by extending hermeneutics, the philosophy of interpretation. Hermeneutics moves beyond the description or core concepts of the experience and seeks meanings that are embedded in everyday occurrences. Heidegger, who was interested in interpreting and describing human experience, believed that bracketing was not necessary because hermeneutics presumed prior understanding. (Polizzi, 2019).

Hermeneutic researchers pursue to reveal aspects of phenomena that are hardly noticed, accounted for, or described. Hermeneutic researchers aim to illuminate important, yet often overlooked, dimensions of human experience in ways that attract attention and provoke more thinking. Hermeneutic phenomenology allows researchers

access to rich contextual data and reveals meaning from human experiences of health care as lived-in and lived-through. It is a methodological method not bound by planned stages of a technique; it is how one accommodates, questions, and thinks in and through evolving methods (Van Manen, 2014). Essential in hermeneutic phenomenology is the place of the researcher's understandings in the interpretive process. The hermeneutic study requires a close understanding of the participants' experiences and the researcher's experience so that a fusion of horizons and closing of personal or historical distance between minds occurs (Crowther, Ironside, Spence, & Smythe, 2017).

Hermeneutic phenomenology is concerned with the exploration of the lifeworld and the study of lived experience or human experience as it is lived. The idea of the lifeworld is basically the world of lived experience. One's lifeworld and world of lived experience is what one experience before one has begun to label or conceptualize it. The concept of the lifeworld derives from the work of Husserl. In his work, Husserl discusses two fundamental ways in which we come to understand the lifeworld: our natural attitude and our reflective attitude (Rich, Graham, Taket, & Shelly, 2013). The focus is toward illuminating details and seemingly trivial aspects within experience that may be taken for granted in our lives, with a goal of creating meaning and achieving a sense of understanding. One's life worlds change depending on the time of the day such as the lived world of home, work, and the lived world of school (Tembo, 2016).

Heidegger developed interpretive phenomenology by extending hermeneutics, the philosophy of interpretation. He expanded hermeneutics by studying the notion of being in the world rather than knowing the world. Hermeneutics moves beyond the description of concepts of the experience and seeks meanings that are implanted in everyday

occurrences. In van Manen hermeneutical phenomenology, research has to do with the lived experience and interpreting the texts of life. Van Manen believes it might be difficult to bracket personal experiences as the assumptions that the researcher brings to the study are always incorporated into the interpretations (Creswell & Poth, 2018).

Husserlian Phenomenology (Descriptive)

Phenomenology can be traced back to the great Greek philosopher, the student of Socrates, Plato (Converse, 2012). It was first identified and described in the works of such philosophers like Immanuel Kant and Georg Wilhelm Friedrich Hegel the early 18th-century. Edmund Husserl, a German philosopher and mathematician, is considered the father of phenomenology who founded descriptive phenomenology (Converse, 2012). Husserl's focus was epistemological; the emphasis was placed on how the object or experience appears to the consciousness. Husserl believed that phenomenology suspended all suppositions, was related to consciousness and was built on the meaning of the individual's experience (Abalos, Rivera, Locsin, & Schoenhofer, 2016). The experience of perception, thought, memory, imagination, and emotion involve what Husserl called intentionality, which is an individual's directed awareness or consciousness of an object or event. This process within Husserl's phenomenology is also known as phenomenological reduction or epoche. Husserl's concepts, epoche, or bracketing in which the researchers put aside their experiences as much as possible and take a new view toward the phenomenon to be studied (Ellis, 2016).

Husserl developed descriptive phenomenology, where experiences were described while preconceived opinions were bracketed. Bracketing or separating out of consciousness what is already known about or believed about the phenomenon being

experienced. This process entails that in order to grasp the essential lived experience of those being studied, the researcher should shed all prior knowledge related to the phenomenon being studied. This technique was not to eliminate the existence of that prior knowledge. It is a process of suspending an individual's judgment or bracketing particular beliefs about the phenomena in order to see it clearly.

Husserl viewed consciousness as a co-constructed discussion between an individual and the world (Shardonofsky et al., (2019).. Furthermore, he saw access to the structures of consciousness not as a matter of generalization, but as a result of direct grasping of a phenomenon. This grasping was seen as an intentional process, actively guided by human intention. Husserl viewed intentionality and essences as key to the understanding of phenomenology.

Heuristic inquiry is a process that starts with a question or problem which the researcher seeks to illuminate or answer. The question is one that has been a personal challenge and puzzlement in the search to understand the world in which one lives. Heuristic research study is a way of engaging in scientific search via methods and processes aimed at discovery, a way of self-inquiry and dialogue with others aimed at finding the underlying meanings of important human experiences (Moustakas, 1990).

Heuristic research comprises self-search, self-dialogue, and self-discovery; the research question and the methodology are out of inner consciousness, meaning, and inspiration. In heuristic research, the researcher must have a direct, individual encounter with the phenomenon being investigated. It requires the total presence, maturity, integrity, and honesty of the researcher who desires to know, understand, and appreciate the phenomenon being studied (Moustakas, 1990). Heuristic research is different from

hermeneutic research in that the focus is entirely and consistently aimed at understanding human experience. Only the co-researchers' experiences with the phenomenon are considered, not how politics, history, arts, or human initiatives explain the meaning of the experience (Moustakas, 1994).

Moustakas' transcendental phenomenology is focused less on the interpretations of the researcher and more on a description of the experiences of participants. Moustakas also focuses on the Husserlian concept of epoche, or bracketing, in which investigators set aside their experiences in order to take a fresh perspective toward the phenomenon under investigation. Descriptive phenomenological studies involve four steps: bracketing, intuiting, analyzing, and describing (Polit & Beck, 2017).

Moustakas' transcendental (empirical) phenomenology is focused less on the interpretations of the researcher and more on a description of the experiences of participants. The empirical phenomenological approach includes a return to experience in order to all-inclusive, full descriptions that provides the foundation for a reflective structure analysis that represents the essences of the experience. When a strictly descriptive approach is adopted, the phenomena are allowed to speak for themselves, and when this happens, one discovers that whatever appears suggests in its appearance that which is concealed. The objective is to determine what an experience means for the individuals who have had the experience and are able to give a comprehensive and complete description of it (Moustakas, 1994).

Relationship of Transcendental Phenomenology to the Study

Information in the literature regarding the experience of sickle cell patients during VOC is limited. A better understanding of the lived experience of adults with SCD who

have experienced VOC is needed; therefore, the phenomenological approach is necessary for this study. Transcendental phenomenology research is descriptive and focuses on the structure of experience, the organizing principles that give form and meaning to the world. According to Moustakas (1994):

> The researcher following a transcendental approach engages in disciplined and systematic efforts to set aside biases regarding the phenomenon being investigated (known as the epoche process) in order to unveil the study as far as possible free of presumptions, beliefs, and knowledge of the phenomenon from previous experience and professional studies—to be completely open, receptive, and naïve in listening to and hearing research participants describe their experience of the phenomenon being investigated (p. 21).

Transcendental phenomenology supports impartiality, where preconceived notions, bias, or judgments are set aside. It is vital to this study that the researcher brackets her prior experience in order to see the experience of each person with SCD who has experienced at lease a vaso-occlusive crisis through his or her own eyes. Rather than an interpretation of the participants' perception of the phenomenon, the researcher is interested in the individual rich descriptions of the phenomenon that made up the essence of the phenomenon being studied. While phenomenology is only one of many qualitative approaches that pursue to understand the lived experience, this method has been selected based on the research question of exploring the lived experience of adults with SCD during VOC.

Significance of the Study

The intended implication of this study was to promote the understanding the lived experience of adults with sickle cell disease during vaso-occlusive crises. Few research has been conducted on this group of patients. The randomness and recurrence of episodes pose a danger to the lives of people with SCD. During crises SCD patients may not be able to give accurate reports of their pain levels; therefore, healthcare providers often question the legitimacy of pain reports of persons with SCD. The findings of this study may increase the healthcare workers' awareness of the need for prompt pain relief during VOC. The findings of this study may contribute to science by revealing information not yet known, thus filling a gap in the knowledge. It may add to the body of knowledge of phenomenological research. People who are not affected by SCD do not have a clear understanding of the experience of what SCD sufferers experience during a VOC. Other disciplines such as psychology, education, and medicine might be interested in the findings, as the care of sickle cell patients is multidisciplinary. The findings from this study may have significance to nursing research, practice, education, and public health policy.

Significance to Nursing

The purpose of this study was to explore the lived experience of adults with sickle cell disease during vaso-occlusive crises. The findings from this research study may have an impact on the present and future status of nursing profession. This study may increase nurses' knowledge about SCD and the approach to managing the disease. Cultural awareness and sensitivity may help minimized the biases that healthcare workers might have about this group of people. Nursing research provides the scientific foundation for

the practice of the profession. This study may help increase the body of scientific knowledge of the profession. There are specific implications for nursing education, nursing practice, nursing research, and health/public policy that will be discussed.

Implications for Nursing Education

The aim of this study was to explore the lived experience of SCD patients during VOC. The findings can be incorporated into Fundamentals of Nursing courses and other practitioner resources to make nursing students more aware of the importance of caring for patients with chronic illnesses. This study can also present the benefits of being culturally sensitive and nonjudgmental when caring for patients. Studying this phenomenon can give nursing educators and students greater insight into the lived experiences of people with SCD in the hope that greater numbers of existing and future nurses will become more educated, equipped and prepared to provide better care in the future.

Implications for Nursing Practice

This study aimed to give voice to SCD sufferers who are sometimes silenced by the negative attitude of some health care providers. This study may help nurses and other healthcare workers understand the incongruity between verbal and nonverbal cues from patients during VOC because of the severity of the pain, and thus help eliminate the assumption that SCD patients are addicted to and seeking pain medication, an assumption that may lead to their being left untreated. The findings from this study may help structure the practice of nursing such that more focus is on providing adequate nursing care to patients regardless of their condition or socioeconomic status. The condemnatory attitudes of health care professionals can be a barrier to the care of SCD patients. There is

a need to reassess and strengthen the cultural sensitivity training and education provided to health care professionals. Culturally sensitive practices can help to bridge the gaps that may exist between one and other people of different experiences. Health care professionals understand that there is increased potential for prolonged admissions, hospitalizations, and readmissions when pain crises are mismanaged. Therefore, it is imperative that nurses consistently make appropriate assessments of patients with SCD to help improve their pain management and overall quality of life. This study can improve nurses' approach to care delivery to patients with SCD.

Implications for Nursing Research

This study's findings may lead to more phenomenological studies being done in all the other chronic illnesses and in different populations. A study like this one can also be replicated in other parts of the nation and globally. This type of study can bring about similar studies on other chronic illnesses or genetic illnesses in the nation and in other parts of the world. Differences and similarities in the experiences of people suffering from fatal chronic illnesses can possibly bring a national and or global change. The findings of this transcendental phenomenology study will contribute to scientific discussions about SCD patients' who experience VOC. The findings of this study may inspire further investigation into how to minimize the suffering of people affected by SCD, prevent the spread of the disease, and reduce the associated mortality rate. More qualitative, phenomenological studies like this one may provide a picture of what is working well and what is not working well. The findings from this study may bring more awareness of phenomenological studies and their contribution to nursing research.

Implication for Health/Public Policy

Currently, patient satisfaction determines many things including reimbursement for hospitalization. The findings of this study may point out the barriers to attaining complete patient satisfaction with every encounter. Healthcare organizations and nurses should develop policies that reinforce the ANA code of ethics and provide compassionate care for SCD patients. Bioethical principles such as respect, beneficence (doing good), and justice (treating people fairly) should be emphasized. The findings of this study may update practice and policy regarding continuous staff education to prepare health care professionals with new ways to teach SCD patients and their families on pain management and coping strategies, thus reducing the frequency VOC. This study can inspire law makers to reinforce effective and adequate pain management. Law makers can develop ways for making sure all healthcare workers abide by pain management guidelines and take pain management more seriously. The findings of this study may provide lawmakers with additional knowledge to inform them about the appropriate and recommended levels of care and services for people living with SCD. Furthermore, better understanding of the studied phenomenon among lawmakers may lead to the implementation of public educational trainings to increase the awareness of SCD.

Scope and Limitations of the Study

The scope of this study comprised of adults with SCD who have experienced at least a vaso-occlusive crisis, living in South Florida. The aim of this study was on exploring the lived experience of this group, thereby giving them voice to express their individual experience during VOC. A limitation of this study was that the researcher is a novice. To overcome this limitation, the researcher relied on her expert committee

members for guidance and advice on maintaining rigor and trustworthiness. Similarly, the study was limited to participants in South Florida only, as a result, results may not be transferable to a larger population. Another limitation was that participants might have been embarrassed, uncomfortable or shy to discuss their experiences during VOC, and not be forthcoming with their responses; they might have divulged to the researcher only the experiences that they were comfortable discussing and sharing with the researcher.

Chapter Summary

This chapter explained and discussed the background of the study. The statement of the problem, purpose, research questions, philosophical underpinnings, and the significance of the study to nursing education, practice, research, health, and public policy were discussed. The purpose of this qualitative phenomenological study was to explore and understand the lived experience of adults with SCD during VOC. The research aimed to give this group of SCD sufferers their individual experience during sickle crises to provide an inductive description of the lived experience, and to gain the understanding of the essence of the experience of during sickle crises. An overview of phenomenology was discussed. The scope and limitations of the study were outlined. Chapter Two will provide a review of the literature.

CHAPTER TWO

Review of the Literature

The purpose of this qualitative, heuristic phenomenological study was to explore the lived experience of adults with sickle cell disease (SDC) during vaso-occlusive crises (VOC). This research aimed to give this group of SCD sufferers a voice to express their individual experience during their vaso-occlusive crises, to provide an inductive description of the phenomenon, and to gain an understanding of the essence of the experience during VOC. A literature review gives the background necessary to sufficiently inform readers so that they understand what research has been done and appreciate the need for the current study of the phenomenon under investigation (Berg & Lune, 2012). According to Moustakas (1994), a literature review is used in preparing to conduct a phenomenological study to assess the research studies relevant to the topic and distinguish designs as well as methodologies in order to obtain new knowledge about the topic. The review of past literature is an exploration of the major concepts with which one is working, introduces and explains them, and provides the motivation for the new research (Berg & Lune, 2012).

An exploration of relevant literature across disciplines was conducted. Different ProQuest search engines provided by Barry University's online library such as Cumulative Index to Nursing and Allied Health Literature (CINAHL), OVID, Dissertation Abstracts, EBSCOhost, Medline, PubMed, and Google Scholar, have shown various studies that have been conducted on sickle hemoglobinopathies, especially SCD. The key words were sickle cell disease, vaso-occlusive crises, health disparities, and pain management. Citations were limited to the English language and by subject to the

exploration of concepts published after the year 2013. Though the researcher intended to work within this time delimitation, there were few other studies found; therefore, the search was updated to include literature published since 2010. The frequent theme across the literature was that there are fatalities, health disparities, and poor pain management associated with sickle cell disease. Four significant content areas were developed from the literature review: historical context, fatalities of sickle cell disease, pain management in sickle cell disease, and health disparities and sickle cell disease. The literature review will begin with the historical context.

Historical context

Long before its acknowledgment in the Western Hemisphere, sickle hemoglobinopathies were known among people of African ancestry. Sickle hemoglobinopathies are inherited disorders of red blood cells. Sickle cell disease was first described in a medical journal by Dr. James Herrick (1861-1954), a Chicago cardiologist, who observed large numbers of elongated sickle-shaped red blood cells in the peripheral blood smear of a twenty-year-old West Indian male with severe anemia (Relling, 2014).

According to Savitt, Smith, Haywood, and Creary (2014), physicians started using the word ‘crisis’ in reference to SCD pain episodes after the fourth case appeared in print in 1922. By that time, Drs. Sydenstricker at the then University of Georgia Medical Department (now called the Medical College of Georgia) and John G. Huck at Johns Hopkins University and Hospital were reviewing and studying numbers of patients with SCD. They and others recognized what came to be known as sickle cell anemia disease as

an illness with its own unique characteristics and applied specific words to describe those characteristics. ‘Crisis’ was one of such descriptors.

In 1923, Dr. Sydenstricker and colleagues wrote that the SCD diagnosis was made without difficulty as the race, the symptoms of anemia, the scleral discoloration, with a history of abdominal crises and rheumatic pains, the presence or history of leg ulcers, all suggest the condition. The diagnosis was made in relation to a specific social context. The social factors that impact the perception of SCD; ancestral disease origin, race, poverty levels, and education levels begin with the diagnosis. By 1923, medical researchers had started to associate SCD with the black race. For decades after, ideas of inferiority about African Americans remained, along with assumptions that specific clinical experiences were characteristic to the black race. Presently, many assumptions about SCD are still made in the medical community. Sickle cell disease and its related painful vaso-occlusive crises are often associated with unpredictability, urgency, bad conduct, narcotic use and abuse (Savitt, Smith, Haywood, & Creary, 2014). The above studies discussed the history of SCD, the pathogenesis, its clinical variability, factors affecting the frequency of VOC, and the complications of SCD.

Fatalities of Sickle Cell Disease

Graves, Hodge, and Jacob (2016) conducted a descriptive correlational study using a longitudinal design with participants completing a web-based electronic diary associated to pain and symptoms during a three-month period. The objective of the study was to examine and address gaps of knowledge concerning variables such as gender, age, and pain as predictors of depression and anxiety for youth with SCD. Participants were obtainned through the Sickle Cell Disease Foundation of California, a community-based

organization that serves approximately 2,000 individuals with SCD in Southern California. The majority of the participants reported about two pain crises per year that necessitated hospitalization. Some participants reported more than three pain episodes per year requiring hospitalization. Descriptive statistics: means, standard deviations, and frequencies were used to describe demographics, anxiety and depression, and quality of life (QOL). Pearson correlations were used to observe the relationships between anxiety, depression, and QOL scores. Bivariate and multivariate analyses were used to observe factors (gender, age, pain, age, and SCD diagnoses) that may have effects on anxiety, depression, and QOL.

The researchers reported, of the 2,194 children and adolescents with SCD in their study, 46% were diagnosed with a depressive disorder; 90% of those with depression had dysrhythmia diagnosed at age nine years, and by age 14 years, the other 10% had a major depressive disorder diagnosed. The cohort with depression experienced more pain and organ damage complications and incurred more health care costs as a result of emergency care. Chronic complications of SCD become more pronounced in adolescence, such as renal and cardiopulmonary dysfunction and avascular necrosis of the shoulders and hips, all of which affect quality of life. The investigators believe that the painful episodic exacerbations of SCD are life threatening and affect the QOL of sickle cell patients. Sickle hemoglobinopathies are one of the world's leading genetic diseases that have severe physical, psychological, and social impacts on both the affected persons and their families. The findings revealed that there is an association between adolescents with SCD who reported more pain frequency, high pain intensity, and increased symptoms of anxiety and depression. The study concluded that there is a relationship between

adolescents and children with SCD and the number and frequency of psychological disorders, especially depression and anxiety.

Lima-Filho et al. (2016) conducted presumed a mixed method study on adult patients with SCD, using stress echocardiography to examine exercise-induced abnormal increase of systolic pulmonary artery pressure (SPAP) in SCD sufferers, which can lead to pulmonary hypertension. Though the study method used was not explicitly stated, however, the study's data analysis discussed both the quantitative and qualitative data, which were analyzed. The objective of the study was to investigate the occurrence of exercise induced abnormal response of SPAP in adult patients with SCD, and to identify the independent predictors of this abnormal response. The sample included 44 adult patients with SCD (22 men, 22 women) who had normal SPAP at rest (tricuspid regurgitant jet flow velocity [TRV] <2.5 m/sec).

Quantitative data were stated as mean, standard deviation values. Qualitative variables were stated as percentage of group or subgroup. Student's t-test compared quantitative variables between the two independent groups. Pearson's chi-square and Fisher's exact tests were used to observe the association between qualitative variables. Correlations between continuous variables were measured using the Spearman coefficient. Some statistically (level of $P < 0.10$) and clinically relevant variables shown by univariate analysis were chosen for multivariate analysis with a logistic regression model, aiming to determine independent predictors of stress-induced pulmonary hypertension. The researchers discussed that pulmonary hypertension is common in patients with SCD with a prevalence of 10–40%. They claimed that pulmonary hypertension occurs related to a poor prognosis, with mortality rates as high as 50%

within two years, often from sudden death, even in mild cases. The investigators further explained that pulmonary hypertension complication is attributed to several mechanisms involving VOC-pulmonary phenomena, pulmonary thromboembolism, in situ thrombosis, recurrent infections, chronic hypoxemia, and hemolysis. The researchers hypothesized that there might be a subgroup of SCD patients with normal SPAP at rest, in whom an abnormal response to increased pulmonary artery flow could be provoked by active exercise, suggesting the occurrence of emerging pulmonary hypertension.

All participants were in a stable state of the disease; last sickling VOC recorded more than two months before the study, and last blood transfusion more than three months before the study. All participants had preserved physical capacity, and mean age of 25 ± seven years (range 18–50 years). All participants were in sinus rhythm, had regional systolic function on echocardiography with normal left ventricular ejection fraction, and no signs of congestive heart failure or significant valve disease. None of the patients had history of systemic hypertension, long term blood transfusion therapy, pulmonary thromboembolism, diabetes mellitus, or others causes of pulmonary hypertension. Determination of normal tricuspid regurgitant jet flow velocity at rest and under exercise in adults with SCD were taken from a control group of 20 healthy individuals matched for gender, age, ethnicity, weight and body surface area, and level of physical activity.

A single investigator performed echocardiography at rest with commercially available equipment. Sickle cell patients and controls underwent a symptom limited exercise stress test, with a treadmill placed alongside the echocardiographic equipment, soon after rest echocardiography. The test was interrupted only when there was physical

fatigue or development of chest pain, dyspnea, sustained arrhythmias, inferior limb claudication, and systemic hypertension or hypotension. When compared with the control group, participants with SCD had resting diastolic blood pressure, hemoglobin, and hematocrit that were significantly lower. There was a substantial enlargement of the four cardiac chambers and an increase in left ventricular mass index, with preserved left and right ventricular systolic functions in the SCD patients as compared to controls. Patients with SCD had cardiac output at rest that was significantly higher than controls. A slightly higher tricuspid regurgitant jet flow velocity and systolic pulmonary artery pressure levels at rest were detected in the group with SCD, as compared to controls.

The researchers reported that, of the 44 patients with SCD, 25 (57%) showed exercise induced increases in systolic pulmonary artery pressure levels. The remaining 19 SCD patients did not show this abnormality, but their creatinine level was significantly increased. There was physiological increase of systolic pulmonary artery pressure with exercise that is related to increased levels of cardiac output, pulmonary flow, and left atrial pressure. Global enlargement of the heart chambers and larger right ventricle dimensions were detected in exercise induced pulmonary hypertensive sickle cell patients. The researchers concluded that SCD might consist of a range of pulmonary vascular abnormalities, ranging from normal systolic pulmonary artery pressure levels both at rest and during stress, to exercise induced abnormal increase in systolic pulmonary artery pressure levels, which may occur in a significant proportion of mainly young adult patients with SCD, during steady state of the disease. This abnormality can lead to the life-threatening pulmonary hypertension at rest that can progress to pulmonary vascular disease and its serious consequences including mortality.

Desai et al. (2017) conducted a quantitative study to compare the pregnancy outcomes among SCD, SCT, and non-sickle cell disease admissions. The study also assessed the risk of adverse pregnancy outcomes for SCD admissions. The type of quantitative study was not mentioned by the author. Data were collected from March 2011 to September 2015 of all maternal admissions in Kasturba Maternity Hospital, Jhagadia, India. The sample size (total delivery admission) was 10,519. Cross tabularization was done to calculate the pregnancy outcomes, diagnosis, morbidities, and treatment procedures by SCD groups. Missing values were excluded from the analysis. Logistic regression was performed to calculate the risk of these pregnancy outcomes.

Each of the pregnancy conditions and outcomes was taken as a dependent variable and sickle cell disease status as an independent variable. Non-sickle cell admission was compared to SCD admission and sickle cell trait admissions. Odds ratios were also estimated comparing SCT with SCD admissions. All of the odds ratios were adjusted for the year of the admissions. The researchers claimed that more than 99% of SCD admissions were anemic. They further explained that the percentage of severely anemic admission was 5.8% among non-SCD admissions and 6.1 among sickle cell trait, compared to 22.1% among SCD admissions. About 9.9% of SCD admissions resulted in stillbirth compared to 4.4% in SCT of the disease and 3.6% in non-SCD admissions. It was also noted that more than half of SCD admissions needed the blood transfusion. The risk of pre-term delivery and Caesarean section was more than three times higher among SCD admissions; about 45.6% of the SCD deliveries were preterm.

The investigators asserted that physiological changes such as the sickling of red blood cells in SCD contributes to microvascular damage that may lead to complications

and affect the growth of the fetus during pregnancy. The study revealed that women with SCD have higher probabilities of abortion, low birth weight, pre-term, and increased stillbirth when compared to non-sickle cell disease pregnancies and deliveries. The authors concluded that the risk of adverse gestation outcomes was significantly higher among SCD admission in comparison with SCT and non-sickle cell disease admissions.

Tolba, El-Shanshory, El-Gamasy, and El-Shehaby (2017) conducted a prospective, randomized, controlled study on 60 children (30 SCD children, 30 healthy children). The aim of the study was to evaluate the right ventricular diastolic and systolic functions by tissue Doppler and speckling tracking imaging in children with SCD. All children participating in the study, ranging from six months to 18 years were divided into two groups. Group A consisted of 30 children with SCD under follow-up at Hematology and Oncology unit of pediatric department, Tanta University Hospital, Egypt. Group B consisted of 30 healthy children as the control group matched in age and sex distribution. Thirty children with SCD and 30 controls were subjected to clinical, laboratory evaluations, and echocardiographic study using GE Vivid 7, including; two-dimensional and tissue Doppler echocardiographic study. All children participating in the study were subjected to careful history taking and full clinical examination including full cardiac examination. Most children with SCD presented with hemolytic attacks and vaso-occlusive crises. Echocardiography was performed, and speckle-tracking imaging was used for evaluation of the right ventricular function.

For quantitative data, the mean and the standard deviation were calculated, comparison between the studied groups was performed with Student's t-test, with $P < 0.05$ was considered statistically significant. Pearson's correlation coefficient was used to

measure correlation between variables. The result showed that children with SCD, compared to healthy children, even at an early age, had impaired right ventricular systolic and diastolic function indices. There were cardiac abnormalities in children with SCD that are secondary to volume overload caused by anemia. Systolic and diastolic dysfunctions were assessed by tissue Doppler echocardiography and speckle tracking imaging. Diastolic dysfunction and pulmonary hypertension have been known to have poor prognosis in patients with SCD.

The investigators believed that impairment of diastolic parameters of right ventricle function might be due to its higher susceptibility related to SCD. The researchers went further to explain that hyperkinetic circulation and vaso-occlusive effects might also have more effect on the right ventricle, which has significantly a smaller mass than the left ventricle, resulting in faster functional derangement. The study concluded by explaining that other factors such as transfusion therapy with iron toxicity and cardiopulmonary injury attributed to VOC effect of SCD on the circulation are involved in the pathogenesis of cardiac injuries in SCD patients.

Gesteira, Bousso, Misko, Ichikawa, and Oliveira (2016) did a study that used integrative review, a comprehensive method that concurrently included experimental and non-experimental research to attain a fuller understanding regarding families of children with SCD by presenting the current state of science and its applicability to nursing practice. The authors independently conducted the survey by using the electronic databases CINAHL (Cumulative Index to Nursing and Allied Health Literature), LILACS (Latin American and Caribbean Health Sciences), MEDLINE (International Literature on Health Sciences), and PUBMED (National Library of Medicine and

National Institutes of Health).

The articles found were numbered in order of location, and the data were organized by the meaning of the information to be extracted from elected posts. The fundamental element of the integrative review is the categorization of studies. In this regard, the procedure is the presentation of results and the descriptive discussion of information through the construction of a summary table containing: a study code according to the classification of the authors; goals; journal; year of publication; study characteristics; and another synoptic table with comparative studies and their levels of evidence. According to evidence-based practice, studies must be evaluated and classified hierarchically according to the level of evidence. In this study the proposal of Melnyk and Fineout-Overholt was adopted for the analysis of the review.

The researchers stated that, “many families of children with SCD face psychosocial challenges related to frequent hospitalizations and restrictions on social activities that trigger impairment to quality of life and psychological symptoms” (p. 284). The study claims that SCD fluctuates family routine, modifies not only the affected individual’s life but also that of their healthy brothers and sisters who experience breaks and interruptions in their routines because of the strains and demands of the affected sibling’s disease. Therefore, there is an imbalance in the stress and psychosocial functioning of family members. These variations in the household are caused by a new lifestyle connected to the care related to constant hospitalizations, specific drug use during times of crises, restrictions on physical activity, the search for a proper diet, and other precautions that are necessary to control the morbidity.

The study discusses that families and children experience several feelings

including suffering and misery caused by the physical and psychosocial burden, which can affect all members, especially the main caregiver. The study further explains that the stress of disease management has consequences on family dynamics, which may lead to a risk of depression in parents. The management of SCD necessitates special care for the sufferers even without them having crises. The intermittent experiences of crises, hospital admissions, and continuous transfusions pose a risk for the psychosocial dysfunction of caregivers and other family members of a SCD patient. The researchers suggested that educating health care professionals is necessary so that the goal of comprehensive care provided to children with SCD and their families is effectively achieved. Additionally, SCD does not affect the individual only but also the family; hemoglobinopathies place a huge emotional, psychological, and economic burden on families. The researchers concluded that SCD is a chronic disease that leads to limitations, despair, loss, and frustration; children fail to attend school when they are in crisis or need hospital care. These physical and psychosocial burdens lead to psychological illnesses that impair the quality of life of both the individuals suffering from the disease and the family members.

Saganuwan (2016) conducted a retrospective study of SCD patients to determine the disease pattern in sickle cell patients from Northwestern Nigeria. Case notes of 319 sickle cell patients were collected and reviewed retrospectively at the Hematology Department, Usmanu Danfodiyo University Teaching Hospital in Nigeria. The review period was between January 1999 and December 2013. Data were organized according to age, sex, weight, and incriminating hematological diseases. The investigators observed clinical signs and their frequencies associated with SCD. There were 89 (45.1%) pain/crises, 56 (28.4%) pneumonia, 18 (9.1%) gastric disorders, 5 (2.5%) musculoskeletal

diseases, 5 (2.5%) renal diseases, 4 (2.0%) priapism, 4 (2.0%) splenomegaly, 1 (0.5%) fever, 1 (0.5%) conjunctivitis, 1 (0.5%) cholecystitis, 3 (1.5%) epistaxis, 1 (0.5%) hemophilia, and 1 (0.5%) acute chest syndrome. Data on SCD and malaria were analyzed using chi-square at 0.1% level of significance. However, data on incidence of SCT were obtained in percentage and their mean hemoglobin values were determined. Analysis of variance (ANOVA) was used to analyze data on weight, red cell volume, packed cell volume, plasma volume, total blood volume, and hemoglobin, and the least significant difference was detected at 5% level.

The researcher observed that pain crises, pneumonia, gastric, central nervous system (CNS), biliary, renal and musculoskeletal disorders, acute chest syndrome, and priapism are all associated with sickle cell VOC. The musculo-skeletal-related abnormalities observed are avascular necrosis of femur head, chronic osteomyelitis of right tibia, acute osteomyelitis, and arthritis while the gastrointestinal disorders observed were gastritis, gastroparesis, and atrophic gastritis and the renal diseases were anuria, urinary tract infection, and polycystitis. The study declared that sickle cell disease could cause conjunctivitis and visual loss in 0.5% of the affected patients, which frequently lead to permanent loss of vision. The investigator also observed disorders related to central nervous system, ischemic heart disease, hypertension, and sleeplessness. The researcher recommended that the use of anti-sickling, anti-inflammatory, analgesic, hematonic, and antimalarial drugs in the treatment of the affected patients might improve the quality of life of the individuals affected by SCD. The study reported that the SCD affected individuals showed significant loss of body weight, anemia, and sickle cell pain crises. In the study, the average survival period for patients with SCD was found to be 49

years.

The above research studies discussed the fatalities of SCD and its associated VOC. They confirmed that SCD painful episodes lead to a significant number of hospital admissions and readmissions each year. The studies explained major complications of VOC such as cardiac injury, depression, anxiety, abortion and pre-term labor. It was also determined that patients with SCD account for the majority of health-care expenses associated with the disease, and the more frequently the visits to hospitals, the more likely the fatality and mortality. However, they did not address what SCD sufferers experience during VOC. This proposed study is about the lived experience of adults with sickle cell disease during vaso-occlusive crises which is different from what these studies have been addressing.

Pain Management in Sickle Cell Disease

Carroll (2015) conducted a quantitative study using correlational descriptive design to measure perceived pain treatment satisfaction and the ability to perform activities of daily living (ADLs). The population of interest was patients with SCD who were experiencing a vaso-occlusive crisis that required an inpatient stay of a minimum of three or more days. The sample consisted of inpatient SCD patients at Seidman Cancer Center in Cleveland, Ohio. Individual subjects completed a demographic form and the revised American Pain Society Patient Outcome Questionnaire (APSPOQ-R) modified for SCD. All surveys were completed with all questions being answered. Data were analyzed using univariate statistics, descriptive statistics, and Pearson's correlation.

The study enlightens that pain affects "116 million people in the United States

and that the healthcare related costs for pain episodes are between $560-635 billion a year due to treatment and the loss in productivity related to painful episodes" (p. 1). Additionally, the study explains that patients with SCD commonly have increased episodes of acute pain, which require treatment in the emergency department, ambulatory clinics, or an inpatient hospital stay.

All patients in the study had some level of severe pain. The participants reported that during the first three days of their admission for pain management, they had severe pain that was 70% on average. About 21% had severe pain 100% of the time, and 42.2% experienced severe pain of 10, on 0-10 scale, 50-60% of the time. The participants described pain relief of only 47% of the time during the first three days of their inpatient admission, on average. The degree of pain relief varied widely with most patients reporting 50% pain relief. Nearly three-fourths reported they did not obtain information about options for their pain relief. The study enlightened that doctors frequently treated SCD pain with decreased amounts of pain medication due to the disbelief in pain reports and the likelihood of addiction and or abuse. The author claimed that SCD patients are often not as pleased with their pain management while in the hospital as compared to other adult population hospitalized for acute pain. The investigator asserted that poor satisfaction was probably due to the sickle cell disease patients not being able to communicate effectively with their providers due to mistrust. The researcher concluded that pain associated with VOC affects the ability to perform daily activities, interferes with the ability to work or fulfill social roles.

Mainous III et al. (2015) conducted a study that was an analysis of a survey conducted as a part of the Council of Academic Family Medicine Educational Research

Alliance (CERA). The survey was done from November 2013, and January 2014, and sent to 3158 doctors who are part of the Council of Academic Family Medicine organizations. The aim of their study was to examine the attitude of family physicians toward SCD management; the comfort level to treat patients with SCD. Data were collected as part of the CERA in the U.S. and Canada that aimed for family physicians who were members of CERA-affiliated organizations. The initial investigators submitted questions related to SCD practice and treatment for inclusion in the CERA survey. The survey questions developed for the study were related to the comfort for managing SCD patients, willingness to manage patients' complication concerns, and usefulness of clinical decision support (CDS) tools.

Comfort with pain management and overall management of SCD patients was evaluated using a Likert scale (very uncomfortable, neutral, and very comfortable). Comfort with managing SCD individuals with specific treatment options (hematopoietic stem cell transplant and red blood cell transfusions, and hydroxyurea) was also assessed. Concern for SCD complications was evaluated using the physician's stated level of concern (very unconcerned, neutral, and very concerned) for identified complications of SCD, including stroke, iron overload, pneumonia, and atherosclerosis. Willingness to co-manage a patient with SCD was assessed for adult and pediatric patients (very likely, neutral, and very unlikely). The willingness of a doctors to self-manage care of SCD patients with the assistance of a CDS tool was also assessed for both adult and pediatric patients (very unlikely, neutral, and very likely).

Data were collected on race/ethnicity, age, primary physician duty, academic rank, time in clinic, patient time, number of patients with SCD, and the proportion of

patients who are African American, and proportion of SCD patients under 19 years of age from all survey participants. Descriptive statistics were computed to understand the general practice patterns of the survey respondents and their overall attitudes toward SCD and SCD treatment. All of the Likert scale questions were condensed into two groups, exploring the difference between those respondents who felt neutral or responded negatively and those who answered the questions with a positive answer. The number of surveys returned was 1060 for a 34% response rate. Bivariate analyses with chi-square tests was conducted to compare attitudes based on respondents' proportion of African American patients (less than 10% versus 10% or greater), as well as by the presence of SCD patients in the physician's practice, and by physician age (younger than 50 versus 50 and older). Statistical significance was judged at $P \leq 0.05$.

There were several significant differences between doctors under age 50 and those aged 50 and older. A smaller percentage of younger doctors were comfortable with management of SCD patients (15.7%) compared to older doctors (25.1%). A larger percentage of doctors who were older expressed concern for iron overload, with 66.1% expressing concern. It was found that academic family doctors had few SCD patients in their patient panel. More significantly, the results of the study show that there are concerns among primary care physicians regarding their ability and skill to manage sickle cell disease, pain crises, and its other complications. Generally, 20.4% of respondents felt comfortable with treatment of SCD. The investigators asserted that there were significant differences in comfort level for treatment of SCD patients depending on whether or not doctors had patients who had SCD, as well as physicians who had more than 10% African American patients. About 69.4% doctors also felt that clinical decision support

(CDS) tools would be useful for treatment and 72.6% felt it would be helpful in avoiding complications in managing SCD patients. The researchers of this study concluded that family physicians are generally uncomfortable with managing SCD patients.

Coleman, Ellis-Caird, John McGowan and Benjamin (2016) conducted a qualitative research design using face-to-face semi-structured interviews on sickle cell pain management. The objective of the study was to provide an in-depth and meaning led account of the experience of SCD pain. Sample included seven adults with SCD (four women, three men), aged 24–57 years, who experienced SCD pain daily or on most days, recruited via an SCD community support group for adults in London, United Kingdom. Interviews were transcribed and analyzed using Interpretative Phenomenological Analysis. Data analysis followed the stages of Interpretative Phenomenological Analysis while exploring the double-hermeneutics or dual interpretation of the data. This involved the participant first making sense of their experience of pain, followed by the researcher decoding this meaning to try and understand what the experience is like from the participant's perspective.

The participants in the study described experiencing unconceivable, excruciating, unceasing, inescapable, unavoidable, and immeasurable pain, which was almost impossible to describe; participants resorted to using analogy and personification as a way to overcome this difficulty. Participants described the pain of SCD as the worst pain that had no cut-off point. In the study, one participant reported that on a scale of one to ten, her pain was a twenty. Some of the participants who had given birth rated SCD pain as much worse. It was found that the pain of SCD negatively affects physical, psychological, and social functioning.

Participants spoke about a process where they felt they had no choice but to accept their disease as it would never be cured; but were able to appreciate life and recognize positive life lessons as a result of living with SCD. Losses associated with living with SCD pain include an inability to exercise autonomy, socialize freely, or fully assume wanted roles. The researchers claim that SCD patients feel their pain experience is misunderstood or not believed. Some participants explained that they had been in an induced coma before and still were in pain. Moreover, many participants thought their pain level was misjudged or not believed because the healthcare providers could not see any physical proof of their pain. The participants further explained that the misunderstanding of their pain caused mistrust and miscommunication with healthcare providers, particularly when extra analgesia as required. Participants described how they were not believed about their pain level, the need for painkillers, and the frustration and annoyance that come along. One participant in the study discussed that though pain is what the patient says it is, unfortunately, the pain of VOC is not considered as such by doctors.

The researchers concluded that sickle cell pain crisis experience is a highly complex and individual phenomenon. Sickle cell patients do not feel that rating scales for pain completely capture their pain experience, but instead the scales leave them feeling misunderstood. Moreover, because current pain measures used in clinical practice appear inadequate at capturing and understanding the acute and chronic SCD pain experience, the researchers suggest that analogies and personification could be used to describe and make sense of the pain of sickle cell vaso-occlusive crises.

Cacciotti, Vaiselbuh, and Romanos-Sirakis (2017) did a single-institution,

retrospective review of medical records to record ED pain management and hospitalization trends. The objective of the study was to review pain scales documentation and the pain treatments used for patients with SCD-related vaso-occlusive pain in the ED. The investigators also aimed to determine factors that affect the type of pain treatment routine prescribed and the amount of time to treatment. The study cohort was chosen using pediatric patients with sickle cell disease, 0 to 21 years old, visiting the ED with an acute pain related complaint, determined by ICD-9–related SCD code, sickle cell with crisis, and acute chest between October 2004 and September 2013. A total of 176 visits were reviewed from 47 patients to record emergency room pain management and hospitalization trends. Variables included demographics, diagnosis, time from triage to initial analgesic medication, analgesic agents, and time to reassessment after initial pain medication was administered. It also includes pain scale at initial presentation and reassessment, ED physician caring for the patient, disposition, and length of stay if hospitalized. Descriptive statistics: mean, median, and standard deviation were calculated on the continuous variables. Frequencies and percentages were organized from the categorical and ordinal variables. The Mann-Whitney or Kruskal-Wallis test, as appropriate, was used to compare groups on a number of chosen continuous variables.

The researchers asserted that most ED visits and hospitalizations for SCD patients are related to painful VOC. Vaso-occlusive crises pain is unpredictable, and the triggers may not be evident. The researchers believed that insufficient pain management in SCD patients is associated with increased morbidity with possible complications in almost all body systems, and that it reduces quality of life in SCD patients of all ages. Among SCD inpatients in the study, 56% received narcotics alone, 27% received Non-Steroidal Anti-

inflammatory Drugs (NSAIDs), and only 17% received combination of NSAID + narcotic as initial treatment in the ED. Average time for pain medication administration was 63 minutes and the average time for pain reassessment after pain medication was 100 minutes. In the study, it was realized that 35% of patients with severe pain, at reassessment in the ED, were already discharged.

Differences exist in the choice of pain management treatments of sickle cell VOC in the emergency department, probably because of treatment by a diversity of ED staff with different methods to pain control. The researchers further enlightened that despite that recommendations indicate that analgesic medications should be provided to VOC patients within 30 minutes of arrival to the hospital, pain medication administration is often delayed; only 20% of adult ED physicians and 9% of pediatric ED physicians report utilizing the recommendations when managing VOC. The researchers concluded that the goal for the initial pain scale assessment in the ED should be to provide quick pain control in SCD patients.

Works, Jones, Grady, and Andemariam (2016) conducted a retrospective chart review of patients with SCD who sought comprehensive medical evaluation at the Sickle Cell Institute at the University of Connecticut Health (UConn Health). This Sickle Cell Institute is the only adult comprehensive SCD program in the northern region of Connecticut, New England. The objective of their study was to examine the relationship between factors such as clinical history, traumatic exposure, and the presence of chronic pain in adult patients with SCD. The sample consisted of 71 SCD patients. The evaluation was integrated by a licensed clinical social worker (LCSW) with expertise in medical social work and behavioral health to identify and address social and emotional

barriers to clinical care.

Pearson's Chi-square test was used to assess associations among categorical variables. When the assumptions of the chi-square test were not met, Fisher's exact test was used instead. For continuous variables that satisfied all of the requisite assumptions, differences between group means were assessed using unpaired two-sample t tests. The ones that did not satisfy the assumption of population normality were instead analyzed using the nonparametric Wilcoxon rank-sum test.

In this study, traumatic experience was defined as perceiving or being threatened with a situation involving actual injury, a threat to the physical integrity of one's self or others, or possible death. Out of the 32 SCD patients who requested mental health counseling, 22 patients identified severe episodes of acute illness, emotional abuse or poor care from medical providers as traumatic. All patients exposed to a traumatic event reported responding to the event with intense fear, horror, helplessness, or emotional shock. In general, there is a high likelihood of exposure to traumatic events for sickle cell patient throughout a lifetime. It appears that patients with SCD may have a higher rate of traumatic exposure than the average population. The higher rate of traumatic exposure in SCD patients may be, in part, due to the nature of SCD, its acute pain crises, as well as the frequency of contact with the medical system, all of which can be traumatic experiences for patients.

The study also confirmed that there was a significant correlation between the presence of chronic pain in SCD patients and traumatic experience, older age, "higher daily pain scores, the need for daily opioids, and greater acute care utilization" (p. 45). The researchers asserted that the longer one lives with SCD, the more likely one is to

have complications related to the disease that may biologically influence the development of chronic pain. The investigators concluded that severe VOC pain episodes are the main characteristic of SCD and the main cause of hospitalizations in SCD patients, and that sickle cell patients are more likely to require hospitalization for VOC pain occurrences, and when admitted, have a longer average hospitalization.

Franceschi et al. (2016) conducted a crossover clinical study between January 2010 and July 2013 on a group of adult patients with SCD referred to the Department of Medicine, University of Verona, Italy. The sample consisted of SCD patients aged 18–45 years with severe painful VOCs, with a visual analog scale pain level (VAS) of 7 or more. The study aimed to assess the effect of fentanyl buccal tablet (FBT) as breakthrough pain medicine in the early phase of pain management during severe VOC in adult patients with SCD. The statistical analysis was performed comparing data from the two treatments of patients with VOC. Nonparametric statistics were implemented. The evaluation of treatment's efficacy, tolerability, and safety and other cumulative calculated measures were performed using the two-tailed Wilcoxon signed-rank test comparing the two treatments. Friedman's nonparametric test for related samples followed by Dunn correction for multiple comparisons was used to compare the time course for each parameter at each interval. A P value < 0.05 was considered statistically significant.

The investigators explained that in SCD, VOC are considered severe constant pain with exacerbation of acute pain. This acute pain is most likely due to abrupt "temporary local enhanced vasoconstriction phenomena that are favored by abnormal parasympathetic vascular response, promoting acute ischemic pain" (p. 686). The acute pain of VOCs is usually managed with increasing or rescue dose of around-the-clock

medications such as oral morphine. The researchers went further to discuss that previous studies in both mouse models and human subjects for SCD showed that treatment with morphine might be associated with renal toxicity, increase rate of acute chest syndrome, and peripheral system dysfunction. In addition, the faster clearance of morphine seen in SCD patients requires the development of alternative or additional beneficial strategies in controlling acute pain of VOC.

The study asserted that pain crisis in SCD is characterized by incessant pain with intensified acute pain related to the dynamic vaso-occlusive phenomena that are negatively affected by vasoconstriction, necessitating a breakthrough pain medication approach. The investigators asserted that fentanyl buccal tablet as a breakthrough pain drug might be a strong and reasonable tool in early management of VOC in EDs as reported in other acute noncancer pains. The authors claim that using FBT as a breakthrough pain medication would significantly improve pain relief during the first phase of severe VOC in adult patients with SCD. The control of acute pain in VOC might reduce more severe sickle-cell-related complications. The researchers concluded that the control of acute pain in the early phase of severe VOCs is still unsatisfactory and negatively affects SCD patients' quality of life.

The above literature review discusses the pain that SCD sufferers endure during VOC and the attitude of ED healthcare workers. The studies showed significant inconsistency in the treatment of sickle cell pain in the ED. They revealed that quick evaluation of pain is critical to ensure speedy pain relief and prevention of complications. The studies suggest that ED workers should provide adequate and appropriate pain management to improve the QOL of SCD patients and prevent complications. However,

the studies did not address the total experience of these patients during VOC, as pain may not be the only issue experienced during VOC. . This proposed study is about the lived experience of adults with sickle cell disease during vaso-occlusive crises which is different from what these studies have been addressing.

Health Disparities and Sickle Cell Disease

Haywood et al. (2014) conducted a quantitative study that involved a cross-sectional analysis of Improving Patient Outcomes with Respect and Trust (IMPORT) participant data. The aim of the study was to evaluate the relationship between perceived discrimination from healthcare providers and non-adherence to physician recommendations among persons with SCD and to test the potentially mediating role of patient trust. Participants were patients with SCD, age 15 years and older. The study used patient self-report items from the 2001 Commonwealth Fund Health Survey; participants completed a comprehensive questionnaire given by an audio computer-assisted self-interview.

For statistical analysis, t-tests and chi-square tests were used as appropriate to examine the bivariate associations among study variables, using differences in means and proportions as our reported bivariate unstandardized measures of effect size. Due to the relatively high frequency of the outcome variable (non-adherence), and because odds ratios from logistic regression are known to overestimate relative risks when the outcome of interest has a high frequency, multivariable Poisson regression models with a robust variance estimator was used to test the independent association of experiences of discrimination with non-adherence to physician recommendations, and to test the role of patient trust as a mediator of the discrimination/non-adherence relationship.

The investigators claimed that participants considered as non-adherent were more likely to report having an experience of discrimination. In the study, the two primary predictors of interest, discrimination and trust, were substantially associated with adherence status at the bivariate level. The researchers reported that among 273 SCD patients with complete data on all variables of interest, 58% of the non-adherent group, compared to 43 % of the adherent group, reported at least one experience of discrimination. However, since 43 % of the adherent group also described having at least one experience of discrimination in the prior two-year period suggests a high underlying degree of perceived discrimination among SCD patients overall.

The results of the statistical analyses of the study provide evidence in support of the researchers' theorized model, signifying that discriminatory experiences in the healthcare setting are related with less SCD patient trust in medical professionals and other healthcare workers, and lower levels of trust in medical professionals are associated with a greater likelihood of non-adherence to doctor's recommendations. The researchers asserted that perceived discrimination appears to affect adherence behaviors through the pathway of patient trust and that discrimination in healthcare is hypothesized as a contributor to the problem of racial and ethnic health and healthcare disparities.

The researchers believe that the experiences of discrimination were associated with significant reductions in trust reported by the patients in their sample. Sickle cell disease patients experience many problems with the interpersonal quality of their care from healthcare providers, and these problems have an impact on the trust that SCD patients have in the medical profession. The researchers concluded that SCD patient perceptions of discriminatory experiences from healthcare providers are associated with

greater non-adherence to physician recommendations and might be a potential factor contributing to disparities in health quality among the SCD patient population.

Carlton, Haywood, Tanabe, Naik, Beach, and Lanzkron (2013) conducted a cross-sectional, comparative analysis of data from the National Hospital Ambulatory Medical Care Survey (NHAMCS) from year 2003 through 2008. The objective of the study was to determine whether or not patients with SCD experience lengthier wait times (in minutes) to see a doctor after arrival to an ED compared to patients with long bone fractures and other patients arriving with all other possible conditions. The study also attempted to separate the effects of race and disease status on any observed differences. National Hospital Ambulatory Medical Care Survey is a nationwide representative sample of nonfederal emergency department visits in the United States.

Data from NHAMCS for the years 2003 through 2008 were used for the study. Emergency Department visit records in the NHAMCS dataset comprise up to three doctor diagnosis codes in ICD-9-CM format. The Emergency Department visit was defined as SCD-related if any one of the three doctor diagnoses had an ICD-9-CM code for SCD. Patients with long bone fractures were selected as a comparison group because both SCD and long bone fracture patients present to EDs with excruciating acute pain necessitating speedy analgesic administration. All other observations were coded as belonging to the general patient sample. Doctor wait times were calculated from time seen by a physician and documented time of arrival to the ED.

Potentially confounding patient-level and hospital-level covariates such as patient sex, age, race, insurance type, assigned triage priority, and recorded pain score at triage, were examined. All statistical analyses used Stata 12.1 software, and chi-square

and T-tests tests were used for all bivariate analyses. A total of 171,789 NHAMCS records met our inclusion criteria. Twenty-two percent of the SCD patients were assigned a triage category of <15 minutes compared to 18.6% of the long bone fracture patients and 16.3% of the General Patient Sample. Likewise, 48.2% of the SCD patients were assigned to the 15 to 60 minute group compared to 39% of the LBF patients and 39.2% of the General Patient Sample. The authors argued that SCD patients were more likely than the long bone fracture and the General Patient Sample patients to present with severe levels of pain, defined as pain scores of 7 to 10. Severe pain at triage was recorded in 54% percent of SCD patients compared to only 32.3% of the long bone fracture patients and 19.4% of the General Patient Sample.

At the bivariate level, SCD patients waited longer to see doctors in the ED than did the long bone fracture patients. The mean wait time for SCD patients was 66.8 minutes compared to 42 minutes for the long bone fracture patients. The difference in mean wait times between the two groups was 24.7 minutes. Likewise, longer ED wait times were observed for SCD patients compared to the General Patient Sample on bivariate analysis. The mean wait time for the General Patient Sample was 53.6 minutes. Sickle cell disease patients waited 13.1 minutes longer, on average, to see physicians in the ED compared to the General Patient Sample.

The researchers discussed that SCD patients experience extended wait times to see a doctor when they arrived at an ED than do other patient populations, despite that the SCD patients tend to express higher levels of pain and tend to be given higher priority triage ratings. The researchers also claimed that the Black race of SCD patients seems to account for much of the variance in wait times between SCD patients and a General

Patient Sample. It was also found that SCD patients experience longer wait times than do patients with long bone fracture. The investigators asserted that both the race of sickle cell patients and their status as SCD patients contribute to delays in receiving timely care in the ED.

The researchers asserted that since the SCD population in the U.S. is predominantly African-American, factors that affect the quality of care received by the general African-American population have the probability to impact most SCD patients in the United States. Even though after removing the effects of race by conducting analyses restricted to African-American ED visits only, it was still found that SCD patients waited about 50% longer to be seen by a doctor in the ED compared to patients with long bone fracture and 25% longer, on average, to see a physician in the ED than did a General Patient Sample.

The researchers concluded that individuals and families with SCD experience lower quality of care than do African-Americans or other minority with other conditions. Prior research has suggested that even among African-Americans, individuals and families with SCD experience lower quality of care than do African-Americans with other conditions. Some ED staff professionals possess bias or negative attitudes about SCD patients. These negative attitudes may contribute to longer delays in the provision of treatment in the ED for patients with SCD.

Xavier de Moraes, Bushatsky, Campos Barros, Ramos Barro, and Bezerra (2017) conducted a cross-sectional, descriptive, quantitative-qualitative study on the vision of patients with SCD on assistance provided in the Family Health Units. The study sample included 26 patients with sickle cell disease volunteers, aged 18 years or older. Other

inclusion criteria included people who were hospitalized in outpatient care or inpatient admission at Hemope Foundation Hospital, living in the State of Pernambuco, Brazil. The investigators pointed out that there more than 27,000 Brazilians with SCD, making this disease to be considered a public health problem in Brazil.

The quantitative analysis was performed using parameters based on relative and absolute measures, and statistical associations of bivariate analysis, as well as graphs and tables constructed with the aid of the Excel 2003 and EPI INFO version 3.5.2 programs with a significance level of 5%. The qualitative analysis was based on the guiding question, “What do you think about the assistance provided by your Health Center?” Where Health Center is referred to the Family Health Unit. The responses of the participants were transcribed in full for the Word 2007 program and analyzed according to the content analysis of Bardin (2009), which allows a qualitative approach through testimonials as raw material.

The researchers enlightened that in 2010, the life expectancy of SCD patients in Brazil was 45 years, which was significantly lower than that estimated for the same individuals living in developed countries. They asserted that SCD is a chronic pathology and that therapy should be based on the prevention of complications and the treatment of symptoms. They encouraged that SCD sufferers should be monitored for growth, somatic development, psychological and organic lesions, as well as specific comorbidities. The researchers suggested that public policies of attention to the SCD patients should be implanted in order to increase their longevity.

In the study, 50% of the participants had no employment relationship, receiving only government benefits, because of the disability from the disease. The investigators

believe that SCD is a chronic pathology that affects all areas of the patient's life; it does not only impact their physical condition, but also their family, social, and work interactions. The researchers concluded that despite policies focusing on SCD, there are still problems in the care of patients with SCD; the lack of knowledge about this pathology and its treatment methods may lead to a doubting of the system by the individual affected by the disease. It was advised that although there are health policies for people with SCD, these alone are not enough; it requires the commitment of health professionals.

Ezenwa et al. (2017) conducted a cross-sectional comparative study using patients from a university-affiliated minority-serving outpatient Comprehensive Sickle Cell Clinic located in Illinois. The aim of the study was to evaluate the pain coping strategies of patients with SCD who experience healthcare injustice from either nurses or doctors during medical visits for pain management. The study sample included 52 outpatient adults with SCD who completed the PAINReportIt Healthcare Justice Questionnaire and Coping Strategies Questionnaire-SCD. Inclusion criteria were diagnosis of SCD, use of opioids for pain crisis, ability to speak and read English, and age 18 years or older. Descriptive statistics such as means, standard deviations, frequencies, and percentages, were computed. Data were analyzed using independent t tests. Statistical significance was set at an alpha of 0.05.

In the study, healthcare injustice was defined as the perception of unfairness of the treatment that a patient experiences from healthcare providers. It is considered important in their care, is associated with adverse psychological and physiological consequences in patients with SCD. Healthcare justice and healthcare injustice groups

were created using a median score of 3.5 as a cut-off. Patients who scored below 3.5 formed the healthcare injustice group while patients who scored above 3.5 comprised the healthcare justice group. The researchers asserted that patients with SCD experience healthcare injustice from both nurses and physicians during medical visits for pain management. The study findings suggest that patients with SCD who reported either healthcare justice or healthcare injustice endorsed different sets of pain coping strategies. It was found that patients with SCD used praying-hoping strategy when they experience injustice. The researchers claim that patients reporting healthcare injustice from physicians used isolation while patients reporting healthcare injustice from nurses used isolation significantly. It was found that patients with SCD used isolation as a pain coping strategy when they experienced injustice.

The researchers concluded that patients with SCD who perceive healthcare injustice from physicians or nurses use different pain coping strategies than patients who perceive healthcare justice from physicians or nurses. They suggested future findings that could inform interventions that decrease healthcare injustice and encourage patients with SCD to use pain coping approaches that are connected with positive health outcomes.

Gomes, Torres, Viana, Pereira, and Caldeira (2014) conducted a qualitative study among 14 health community agents from a municipality with a high occurrence of SCD in Brazil. The objective of the study was to determine health community agents' opinions on access and care delivery to individuals with SCD. In the study, 14 patients with SCD were chosen from the records of the municipality's Neonatal Screening Program, made obtainable by the Center for Actions and Research in Diagnostic Support. This qualitative study was done in the city of Janauba, north of Minas Gerais, in access and care of

individuals with SCD in a primary care service in the southeast region of Brazil. The relationship and rapport between each patient and the family health unit in which he/she was registered was investigated. Health community agents from Family Health Strategy teams whose coverage area included patients with SCD were invited. Data collected from participants and observers' speeches were submitted to content analysis in the modality of thematic analysis. Data were organized by interpretative categories.

The investigators claimed that parents of SCD children reported having to wait for assistance numerous times unless the child had a fever or felt very ill. The professionals' lack of knowledge concerning prioritization of care and consciousness of alert signs for possibly severe events was evident. In the study, it was found that professionals did not know the exact care required during follow-up of patients with SCD, therefore, quality of care seemed compromised. Results of the study showed that the healthcare team did not feel responsible for organizing or coordinating care delivery to SCD patients; they felt that the patient's family was responsible for seeking the best care. The researchers asserted that persons with SCD have difficulty accessing primary care because of some special issues such as having a genetic disorder probably because the population of people with SCD is predominantly black. They found that special vaccine coverage was deficient; children with SCD had an incomplete immunization schedule. The study concluded healthcare delivery for patients with SCD, based on health community agents' opinion, was insufficient. It showed that professionals did not know the particularities of follow-up necessary for patients with SCD, and the patients had limited access to the basic health unit.

The above literature clearly confirms that the pain experienced during crises is either undertreated or inappropriately managed because of the low attention paid by healthcare providers to these patients during their sickle crisis. Understanding the lived experience of patients with SCD during VOC will help frontline health care workers provide adequate pain management for SCD patients without prejudice or biases.

Experiential Context

Some of my siblings and I have sickle cell trait but not sickle cell disease. However, I have extended family members, friends, and students who suffer from sickle cell disease and have experienced at least one vaso-occlusive crisis. I also have a close friend whose daughter died from complications of SCD at five years of age. I have supported and observed few friends during their vaso-occlusive crises. I had also taken care of many sickle cell patients when I worked in the acute care setting. I never thought I would be researching this topic of sickle hemoglobinopathy. I intended to research a phenomenon in teaching. Then I thought I should probably research the experience during VOC, both in terms of pain and psychological issues. My experience taking care of SCD patients and having some of them as friends and as students made me develop a passion to study the hemoglobinopathy disease, sickle cell disease.

With my passion for sickle hemoglobinopathy, I bracketed my experience as a family, friend, and teacher of the people who suffer from SCD in order to truly hear and grasp the experiences of my participants. In order to achieve this, I relied heavily on reflexive journaling, both before participant interviews and afterward. Polit and Beck (2017) provided ten steps to help qualitative researchers with bracketing in a reflexive journal:

1. Make note of interests that, as a researcher, one may take for granted.
2. Clarify your personal values and identify areas in which you know you are biased.
3. Identify areas of possible role conflict.
4. Recognize gatekeepers' interest and make note of the degree to which they are favorably or unfavorably disposed toward your research.
5. Identify any feelings you may have that may indicate a lack of neutrality.
6. Describe new or surprising findings in collecting and analyzing data.
7. Reflect on and profit from methodological problems that occur during your research.
8. After data analysis is complete, reflect on how you write up your findings.
9. Reflect on whether the literature review is truly supporting your findings, or whether it is expressing the similar cultural background you have.
10. Consider whether you can address any bias in your data collection or analysis by interviewing a participant a second time or reanalyzing the transcript in question. (pp. 471-472).

Chapter Summary

This chapter discussed a review of the literature. Starting with a historical background of sickle hemoglobinopathy, this chapter discussed and synthesized content areas of fatalities of sickle cell disease, pain management in sickle cell disease, and health disparities and sickle cell disease. This chapter concluded with a description of the researcher's experiential context. Chapter Three will provide a discussion of the methods.

CHAPTER THREE

METHODS

The purpose of this qualitative, Husserl's descriptive phenomenological study was to explore the lived experience of SCD sufferers during VOC. Selecting a method of choice was guided by the research question and the purpose of the study. The research question focused on what it is like to experience a phenomenon, therefore, a qualitative approach will be necessary. The chosen method for this study was the transcendental phenomenological perspective of Moustakas.

Research Design

The research design provides the framework of the study. Descriptive phenomenological studies involve four steps: bracketing, intuiting, analyzing, and describing (Polit & Beck, 2017). Moustakas' transcendental phenomenology is focused less on the interpretations of the researcher and more on a description of the experiences of participants. Moustakas (2012) described transcendental phenomenology as having three core processes: epoche, transcendental-phenomenological reduction, and imaginative variation. Epoche is a Husserlian concept in which the everyday understandings, judgments, and knowing are set aside. Moustakas' approach is systematic. The steps are: (1) the researcher determines if the research problem is best examined using a phenomenological approach; (2) The researcher identifies a phenomenon of interest to study; (3) The broad philosophical assumptions of phenomenology are recognized and specified by the researcher; (4) Data are collected from the individuals who have experienced the phenomenon. Ultimately, the researcher has to build on the data and highlight "significant statements", sentences, or quotes that

provide an understanding of how the participants experienced the phenomenon (Creswell & Poth, 2018, p.471).

A textural description of the essence and meanings of the phenomenon is derived from the transcendental-phenomenological reduction. Descriptive or transcendental phenomenology was used to address the lived experience of persons with SCD during VOC because Moustakas' transcendental phenomenology allows for full expression of the participants' thoughts, feelings, and perspectives as it relates to the phenomenon.

According to Moustakas (1994), the Moustakas' approach is systematic, and the procedural steps are:

1. The researcher determines if the research problem is best examined using a phenomenological approach. The type of problem best suited is one in which it is important to understand several individuals' common or shared experiences. These shared experiences could be used to develop a deeper understanding about the features of the phenomenon.
2. A phenomenon of interest to study is identified.
3. The researcher recognizes and specifies the broad philosophical assumptions of phenomenology. To fully describe how participants view the phenomenon, researchers must bracket out, as much as possible, their own experiences.
4. Data are collected from the individuals who have experienced the phenomenon. Often data collection involves in-depth and multiple

interviews with participants.

5. The participants are asked two broad general questions: What have you experienced in terms of the phenomenon? What contexts or situations have typically influenced or affected your experiences of the phenomenon? Other open-ended questions may also be asked.

6. Building on the data from the first and second research questions, data analysts go through the data and highlight "significant statements," sentences, or quotes that provide an understanding of how the participants experienced the phenomenon. Moustakas called this horizontalization. Then, the researcher develops "clusters of meaning" from these significant statements into themes.

7. The significant statements and theme are then used to write a description of what participants experienced, called textural description. The statements and themes are also used to write a description of the context or setting that influenced how the participants experienced the phenomenon, called structural description. Moustakas has an added step that involves researchers writing about their own experiences.

8. In the last step, the researcher writes a composite description from the structural and textural descriptions that present the "essence" of the phenomenon. It is a descriptive passage, and the reader should come away from the phenomenology with the feeling of understanding better what it is like for someone to experience the given phenomenon (p. 81-82)

The research question of this study was: What is the lived experience of adults with sickle cell disease during vaso-occlusive crises? Quantitative studies have listed various attributes; however, a list of factors without explanation or understanding is not adequate to get a complete picture. A qualitative approach was suitable because not much is known about this phenomenon. Specific to phenomenology, the essence of the lived experience of adults with SCD during VOC. There was a need for a qualitative study in order to hear from these sickle cell disease sufferers in their own words.

Sampling/Setting

Selecting a small portion of the population that accurately represents the target population is the purpose of sampling. One general guideline for sample size in qualitative research is not only to study few individuals but also to collect extensive detail about the individuals studied (Creswell & Poth, 2018). Purposive sampling was used in this study to select study participants who have experienced the phenomenon; they can purposefully inform an understanding of the research problem (Creswell, 2013). Participants who have experienced the phenomenon were informed of research interest. Snowball sampling was also be utilized, which involved asking participants to refer other participants.

When selecting sample size in qualitative studies, the guiding principle is data saturation. There are no fixed rules for sample size in qualitative research (Polit & Beck, 2018). Phenomenologists usually rely on very small samples. Creswell and Poth (2018) discussed that in phenomenology, participants have ranged from “1 to 325” (p. 159). The sample for this study included 20 adults with SCD in South Florida. Sample size in qualitative research depends on several factors. Sample size should be based on

informational needs; therefore, the guiding principle is data saturation. Data saturation is sampling to the point where no new information is achieved, and redundancy is realized (Polit & Beck, 2018). The setting for this study was South Florida. Data collection took place in South Florida in a safe and quiet location mutually agreed upon by the researcher and each participant.

Access and Recruitment of the Sample

After approval from Barry's Institutional Review Board, access to adults with SCD who had experienced at least one vaso-occlusive crisis were sought from the following sources: network of nurses, snowball sampling, social networks such as Facebook and Instagram. Flyers (Appendix C) were given to participants seeking their participation, and also asking them to give flyers to individuals they thought might want to participate. Flyers were posted on Facebook and Instagram inviting people who were interested to participate.

The following recruitment measures were utilized:

1. Flyers were given and sent to friends and family
2. Snowball sampling
3. Social media such as Facebook and Instagram

The flyer included the researcher's contact information where interested respondents learned how to participate in the study. The flyer listed the purpose of the study, inclusion criteria, and the token of appreciation of a $25 American Express gift card. Snowball sampling was utilized by asking participants who agreed to partake to identify other people with SCD who might also be interested in taking part in the study.

A face-to-face interview was scheduled for everyone who met the inclusion criteria and expressed interest in participating in the study. The researcher and the participants mutually identified where the interview would take place. A safe location such as a hospital, university campus, coffee shop conference room, or the participant's home was determined. A mutually agreed upon time was decided upon for the interviews.

Inclusion Criteria

Participants were adults people aged 18-65 years, with SCD, who had experienced at least one episode of vaso-occlusive crisis, living in various South Florida counties (i.e., Broward, Miami, Palm Beach). Other inclusion criteria included English fluency, the willingness to speak openly about their experience and be digitally recorded.

Exclusion Criteria

Exclusion criteria included participants who did not have SCD. Being under 18 years or more than 65 years was an exclusion criterion. Noticeable limited English proficiency and an unwillingness to speak about the VOC experience were additional exclusion criteria. Non-South Florida residents was excluded.

Ethical Considerations/Protection of Human Subjects

All scientific inquiry involving human participants must address ethical issues, and researchers must respect the rights of their research subjects. This study was conducted in an ethical manner to ensure the rights and privacy of all research participants. Approval from the Barry University Institutional Review Board was obtained prior to accessing participants, recruiting participants, and collecting data. Barry's ethical consideration standards was maintained throughout the study. Participants who responded to the flyer were contacted through telephone by the researcher. Once

interested participants contacted the researcher, information about the study was provided to them. Those who met the inclusion criteria and expressed interest in participating in the study were recruited, and a mutually agreed upon time, and a safe place were set for data collection. The study participants were informed that participating in the study was voluntary and that they could terminate participation at any time with no repercussion.

Participants were given the opportunity to select a pseudonym for the study that was recorded on the informed consent. The pseudonym was used on the demographic questionnaire, the digital recordings and the transcript of the recordings. Only the researcher knew the pseudonym, and any statements published in the final report or subsequent publications were reported using participant pseudonyms. Transcription of the data was done by a transcriptionist who signed a third-party confidentiality agreement. Confidentiality was assured through the restriction of access to electronic information on the principal investigator's password-protected personal computer; data and consent were kept in separate files. Additionally, there is restriction of access to hard copy data as they are kept in a separate locked file cabinet from the consent in the researcher's home office. Data will be kept a minimum of five years from completion of the study and indefinitely afterward.

Obtaining informed consent is important for participants' right to privacy and respect. Participants were informed that there were no known risks or benefits to them regarding their contribution to this study. The informed consent provided an understanding of the study and served as permission to conduct the audio recording of face-to-face interviews. The informed consent was collected in person for the face-to-face interviews. Also, the informed consent was kept separately from the demographic form

and other interview data. The audio recording started only after the participant's pseudonym had been chosen and permission had been granted. The audio recordings were destroyed once member checking was completed. Only the researcher knew the pseudonym, and any statements published in the final report or subsequent publications were reported using the participants' pseudonyms.

Participants were fully informed that the study was voluntary and that withdrawing from participation was acceptable at any time without any negative repercussions. They were told they had the right not to answer any research questions that they did not want to answer at any time during the interview. They were told that they had the right to request that audiotaping be suspended at any point during the interview. They also were told a $25 American Express gift card would be given to each participant to keep as a token of appreciation even if they withdrew from the study.

Data Collection Procedure

Data collection began after approval from Barry University's IRB. The methodology used in phenomenology is different than most other research methodologies because the objective is to describe a lived experience, rather than explaining or quantifying it. Phenomenology makes use of a variety of methods including conversations, interviews, participant observation, focus meetings, action research, analysis of diaries and other personal texts. Interviews and participants observation were primarily used in this study. Interviews were less structured, but they were more open-ended to encourage the participants to share details regarding their experience during VOC. Each interview was a maximum of 60 minutes in duration.

The researcher and the participants determined where the interview took place. A mutually agreed upon time was decided for interviews, and a safe location was determined. The researcher utilized her personal cell phone and her Barry University email address as resources for communicating with study participants. Interviews were recorded using two digital recorders and transcribed verbatim after each interview. The use of two audio recordings was to guard against equipment malfunctioning. The researcher explained the purpose of the research study and how the responses would be used and the degree of how confidentiality and rights would be protected. Study participants had the right to withdraw from the study at any time as promised. They also had the right not to answer any research question when they decided not to and the right to demand that audiotaping be terminated at any point during the interview.

An opportunity was provided for the participants to voice any concerns and ask any questions that they might have concerning the study. The researcher addressed any fears or concerns and answered any questions before obtaining the signed consent, which gave the researcher authorization to interview them and permit the use of their individual responses in the study. After each participant had signed the consent (Appendix B), the $25 American Express gift card was given to the participants to thank them for their participation. They were informed that the gift was for them to keep, even if they chose to withdraw from the study. Next, the participants were asked to provide a pseudonym that was used to protect their identity. The participants were then asked to complete a demographic form (appendix E) using their pseudonym. They were also informed that the demographic form would take approximately 10 minutes to complete.

The participants were informed about the purpose of the study, the meaning and essence of informed consent, the risks and benefits, as well as the process for recording the interview. They were informed that there were no benefits or risks for participating in the study. The participants were advised that strict confidentiality would be maintained for the audio recordings and any notes that might have been taken during the interview. Furthermore, a transcriptionist, who signed a third-party agreement, transcribed digital audio recordings. The participants were told that they could stop the interview at any time, and they could also refuse to answer a question(s) or ask that the recorder be turned off or suspended at any time during the interview. Likewise, the participants were informed that the data collected would be used for the study, unless they withdrew from the study.

The format was semi-structured with open-ended questions. The interview questions developed by the researcher were used as a guide. Permission to start the audiotape at the beginning of the interview was requested. At the end of the interview, the researcher thanked the participant and reminded him or her that privacy by the researcher would be maintained. The participants were informed that data would be stored, transcribed, and used for the study. The audio recordings were destroyed after the initial member check was completed. After the first interview, the researcher journaled all observations made in addition to noting any verbal and nonverbal communication as well as the participants' body language. Moreover, the researcher indulged in self-reflection of the process of the interview. Upon conclusion of the interview, participants were thanked for their participation.

After each interview, data was transcribed verbatim by the transcriptionist (Appendix F), and after being reviewed by the researcher, the transcription was emailed to the participants. A 20-minute telephone call was scheduled with the participant to review the transcript for accurateness. The phone call was not recorded; it was just for member checking and to enhance credibility. Member checking is a method of confirming the data through debriefing and discussion with the participants. Data were collected until saturation was met. The transcribed data would be kept for a minimum of 5 years from completion of the study and indeterminately thereafter. The researcher decided the date and time for the member check interviews via telephone call, which was not more than 20 minutes each. The total time for participation in this study was 90 minutes.

Interview Questions

Most qualitative data are collected by interviews instead of questionnaires (Appendix D). Researchers who have no preconceived view of content may conduct unstructured interviews (Polit & Beck, 2017). Researchers may sometimes want to be sure that a precise set of topics is covered in their qualitative interviews, so, they use semi-structured interviews. In semi-structured interviews, researchers may prepare a written topic guide, which is a list of areas to be covered with each participant (Polit & Beck, 2017). This method enables that the researcher obtains all the essential information and gives the participants the liberty to provide as many illustrations and descriptions as they wish. According to Polit and Beck (2017), in preparing the topic guide, “questions should be ordered in a logical sequence……., perhaps from general to the specific” (p. 510).

The focus of this study was the lived experience of adults with sickle cell during vaso-occlusive crises. The purpose of qualitative interviews was to converse with participants and obtain as much information as possible from them regarding the phenomenon in this study. Therefore, open-ended questions were used to encourage meaningful discussion and allow the participant to speak freely. The primary question was “what has it been like for you to experience vaso-occlusive crises?” Additional probing questions were used as needed to provoke additional description and details based on the participant's responses.

Demographic Data

The researcher developed a demographic form (Appendix E). Participants completed the demographic form using their pseudonym after the consent form had been signed. Data on the demographic form was collected for the purpose of describing the sample. Completion of the demographic form did not take more than five minutes.

Data Analysis

The process of analyzing data involves the researcher becoming fully immersed in the rich, descriptive data and using processes such as coding and categorizing to organize the data. The goal is to develop themes that can be used to describe the experience from the perspective of those that lived it. After collecting data through interviews with the participants who have experienced the phenomenon, the data will be analyzed using Moustakas’ (1994) transcendental phenomenological data analysis steps. The data analysis begins with bracketing or the epoche process where the researcher recognizes or reads a subjectivity statement of personal experiences and sets aside preconceptions and biases about the phenomenon being explored (Moustakas, 1994). The full process

includes preparing data for the analyses, reducing the data phenomenologically, engaging in imaginative variation, and uncovering the meaning of the participants' experience.

Moustakas elucidated that the procedures for summary and outcomes should include relating the study findings to and distinguishing it from findings in the literature review. This researcher used the following data analysis processes identified by Moustakas (1994) found below and in figure 3:

1. Listing and Preliminary Grouping
    a. List every expression relevant to the experience (horizontalization).
2. Reduction and Elimination: Determine the Invariant Constituents:

    Test each expression for two requirements:

    a. Does it contain a moment of the experience that is necessary and sufficient for understanding it?
    b. Is it possible to abstract and label it?

    If so, it is a horizon of the experience. Overlapping, repetitive, or vague comments are removed. Expressions not meeting the above requirements are eliminated or presented in more exact descriptive terms. The horizons that remain are the invariant constituents of the experience.
3. Clustering and Thematizing the Invariant Constituents:
4. Cluster the invariant constituents of the experience that are related to a thematic label. The clustered and labeled constituents are the core themes of the experience.
5. Final Identification of the Invariant Constituents and Themes by Application: Validation. Check the invariant constituents and their

accompanying theme against the complete record of the research participant.

   a. Are they expressed explicitly in the complete transcription?
   b. Are they compatible if not explicitly expressed?
   c. If they are not explicit or compatible, they are not relevant to the co-researcher's experience and should be deleted.

6. Using the relevant, validated invariant constituents and themes, construct for each co-researcher an Individual Textural Description of the experience. Include verbatim examples from the transcribed interview.
7. Construct for each co-researcher an Individual Structural Description of the experience based on the Individual Textural Description and Imaginative Variation.
8. Construct for each research participant a Textural-Structural Description of the meanings and essences of the experience, incorporating the invariant constituents and themes.

From the Individual Textural-Structural Descriptions, develop a Composite Description of the meanings and essences of the experience, representing the group as a whole (Moustakas, 1994, p. 120-121).

Moustakas' Data Analysis Approach

Figure 3. Adapted from Phenomenological Research Methods, by Moustakas, C. (1994). Thousand Oaks, CA: SAGE Publications, Inc.

Rigor and Trustworthiness

Rigor of a scientific study refers to the strictness and severity of the study. Quality is known or verified by the rigor of a qualitative study. Trustworthiness is a way of establishing or ensuring the quality of a study. Credibility, dependability, confirmability, and transferability describe the techniques to ensure trustworthiness of qualitative inquiry (Lincoln & Guba, 1994). Qualitative researchers justify research rigor by the trustworthiness of the research, which is when the findings accurately mirror the meanings as described by the participants. This study wss based within the assumptions and characteristics of the hermeneutic phenomenological approach. Maintaining rigor in this study was simplified by accurately and precisely representing participants' experiences.

Credibility

Credibility refers to the confidence in the truth of the data and interpretation of them (Polit & Beck, 2017). Credibility involves the establishment that a research study is believable. A technique to ensure credibility is member checking. Member checking

involves participants reviewing transcripts for accuracy. Continuing data collection until saturation is achieved is another way to enhance credibility. Credibility involves the establishment that a research study is believable. Contrary to quantitative research which depends on the statistical data collected, credibility is dependent on the accuracy of the information collected. Any concern with credibility should be validated by member checks which is the most critical method for establishing credibility (Seale, 1999, p. 468). To maintain credibility, a transcript copy was given to each participant to verify the accuracy and correctness of the description of their words and enable them to provide feedback regarding the translation of the data.

Dependability

Dependability means to the stability of data over time and situations (Polit & Beck, 2017). Dependability ensures that the research results are dependable, and could be repeated. Dependability is the extent that the study could be repeated by other researchers and that the findings would be consistent (Lincoln & Guba, 1985). Audit trails and using rich description will be used to provide dependability in this study. Dependability ensures that the research results are dependable, and could be repeated. It is measured by the way and standard the research study is conducted, analyzed, and presented. Every step, method, and procedure in the study was well explained and stated to allow other researchers to repeat the study and reach the same conclusion, and get comparable and similar results. There can be no dependability without credibility (Lincoln & Guba, 1994); therefore, dependability is demonstrated with the credibility of the findings. In the inquiry of the lived experience of adults with SCD during VOC, the sampling procedure

and data collection procedure were described in detail so that others could read it, assess the method, and regard the study to be useful.

Confirmability

Confirmability is the degree of neutrality in the research study's findings (Lincoln & Guba, 1985). Confirmability can be established by an audit trail and reflexivity. Bracketing and maintaining a reflexive journal will be used in this study. Confirmability questions how the research outcomes are supported by the information gathered. It refers to the impartiality of the study. Confirmability is a process that establishes whether the researcher has been biased during the study. Other researchers can confirm if a research study is mostly based on the researcher's views and biases. However, it is important that researcher bias does not alter the result. Bracketing was utilized to assist the researcher to avoid any preconceived ideas regarding the lived experience of adults living with SCD during VOC.

Transferability

Transferability is the extent to which findings can be transferred to other settings (Polit & Beck, 2017). Excellent and rich descriptions enrich transferability of a study. Rich description is achieved when the researcher collects sufficiently detailed descriptions of data and reports them with sufficient detail to allow decisions about transferability to be made by the reader. Transferability is the degree to which a research study can be transferred to other circumstances or situations. When a research situation and approaches can be related with similar situations that the reader is conversant with, it is said to be transferrable. Transferability was attained by providing a thorough

description of this study from purposive sampling to data analysis. This process would provide the opportunity for the study to be replicated or allow for transferable results.

Chapter Summary

This chapter discussed the methods for this research study. The sample and setting including how the sample will be accessed and recruited was addressed as well as inclusion and exclusion criteria for the participants. Ethical considerations for this study were provided. Data collection and data analysis techniques were discussed including the interview questions, demographic data created, and principles central to research rigor.

CHAPTER FOUR

FINDINGS OF THE INQUIRY

The purpose of this qualitative, heuristic phenomenological study was to explore the lived experience of adults with sickle cell disease during vaso-occlusive crises. Twenty participants were interviewed over four weeks, beginning on July 18, 2019, and ending on August 13, 2019. Data saturation was achieved after interviewing 20 participants. This chapter includes demographic information of the participants, characteristics of the participants, themes, individual textural and structural descriptions, composite textural and structural descriptions, and an overall synthesis of the essence of the experience. A connection of the themes to the theory of Self-care of Chronic Illness is also presented.

Sample Description

Purposeful sampling was used to choose participants who have experienced the phenomenon. In addition, snowball sampling was used, which involved asking participants to refer other participants who they believed may have experienced the phenomenon. The researcher recruited and interviewed participants until a variation of different experiences and data saturation was achieved. The sample size in this study was 20 South Floridian adults, with a diagnosis of sickle cell disease, who have experienced at least a vaso-occlusive crisis. The selection of participants for this study was based on the following inclusion criteria: adult people aged 18-65 years, with SCD, who have experienced at least a vaso-occlusive crisis, living in various South Florida counties (i.e., Broward, Miami, Palm Beach). Other inclusion criteria included English fluency, the willingness to speak openly about their experience and be digitally recorded. All

participants who met the inclusion criteria and signed an informed consent were interviewed. Individuals who did not fall within the age category and did not live in South Florida were excluded. Further, people who were not fluent in English or willing to speak openly about their experience were excluded.

Participants indicated that they have experienced a sickle cell crisis at least few times a year. Some participants had frequent VOC while other participants only experienced a few episodes yearly. Some participants are incapacitated by the disease and the frequent vaso-occlusive crises while others are still active even with the disease. Some are able to maintain their jobs, while others are unemployed because of the frequency of their VOC. Most participants reported having between one to ten VOC per year while two participants reported having at least 30 and maximum of 60 VOC per year.

Demographic Representation

This study is comprised of a diverse group of participants who have experienced at least one vaso-occlusive crisis in different counties of south Florida. The demographic information shown below in Table 1 summarizes the demographic data that was collected.

Table 1

Demographic Characteristics of Participants (*N = 20*)

Age Group	Gender	Race/ Ethnicity	Educational Level	Occupation	Crises/Year
18-30	M	African American	College Student	Server	2-3
18-30	M	African American	College Student	Retail Worker	1-2
18-30	F	Jamaican	Bachelor's Degree	Nursing	7-8
18-30	F	Jamaican	High School	Unemployed	9-10
18-30	M	Bahamian	High School	Unemployed	5-6
18-30	F	African American	Master's Degree	Business	1-2
18-30	F	African American	Associate Degree	Unemployed	9-10
18-30	F	Grenadian	High School	Unemployed	1-2
18-30	M	African American	High School	Unemployed	40-60
18-30	F	African American	Associate Degree	Unemployed	30-40
18-30	F	African American	Bachelor's Degree	Non-professional	5-6
18-30	F	African American	Associate Degree	Non-professional	9-10
18-30	M	African American	College Sophomore	Sales Person	5-6
18-30	M	African American	Nursing Student	Unemployed	5-6
31-40	F	African American	Bachelor's Degree	Professional	5-6

31-40	F	African American	Master's Degree	Professional	5-6
31-40	M	Bahamian	High School	Unemployed	7-8
31-40	F	African American	Associate Degree	Home-care	9-10
31-40	F	Grenadian	High School	Unemployed	1-2
51-60	F	Jamaican	Bachelor's Degree	Unemployed	9-10

Table 1 represents the demographic characteristics of the sample participants, including age, gender, race, level education, occupation, and number of crises per year. The sample size included 20 persons, living in South Florida who had a diagnosis of sickle cell disease who have experienced at least a vaso-occlusive crisis in a lifetime.

Seventy percent (*n = 14*) of the participants were between the ages of 18-30, 25% were between ages 31-40 (*n= 5*) and 5% (*n* = 1) was in the 51-60 age old group. None of the participants were in the 61 and over age group. Sixty-five percent *(n=13)* of the sample population were females, and 35% *(n=7)* were males. Seventy percent of the participants were African Americans *(n=14),* 15% were Jamaicans *(n=3),* 10% were Bahamians *(n=2),* and 5% was Grenadian *(n=1).* Highest education degree levels were as follows: 10% (*n* = 2) hold master's degree, 20% (*n* = 4) hold bachelor's degree, 20% (*n* = 4) hold associate degree, 25% *(n=5)* have high school diploma, and 25% *(n=5)* were college students.

Twenty percent (*n = 4*) of the participants experience VOC 1-2 times per year, 5% (*n=1*) experience VOC 2-3 times per year, and 30% (*n=6*) experience VOC 5-6 times per year. Ten percent (*n=2*) experience VOC 7-8 times per year, 25% (*n=5*) experience

9-10 times per year. Five percent (*n=1*) experience VOC 30-40 times per year, 5% (*n=1*) experience VOC 40-60 times per year, and none of the participants experience declared having VOC 3-4 times per year. Forty percent (*n=8*) of the participants were unemployed because of the frequency of their VOC, 35% (*n=7*) maintain non-professional jobs with some level of difficulty, 15% (*n=3*) have professional jobs with some difficulty, and 10% (*n=2*) are business owners who report being more stressed related to their SCD status.

Characteristics of Participants

Confidentiality was maintained as all participants selected pseudonyms to conceal their identities. They are all sickle cell disease patients living in Broward, Dade, and Palm Beach counties. Participants were selected on a voluntary basis, and consent for participating in the study was obtained prior to starting the face-to-face audio-recorded interviews. The following descriptions of the participants were obtained from the information provided on the demographic form and the audio-recorded interviews.

Brad is a 20-year old college student, a sophomore at a university who reported having crises about one to two times per year. He explained that he had more VOC as a child that has gotten better over the years. He explained that hydroxyurea, an antineoplastic drug, had helped to reduce the frequency of his VOC significantly.

Cerena is a 26-year-old, with SCD who considers the disease as isolating and depressing because of its genetic and chronic nature. **Cerena** declared that she has been able to manage her SCD pain and get it more under control by using different kinds of alternative therapies. She explained that before she started using these therapies, she had a daily issue with severe pain and physical limitations.

Chris is a 20-year-old college student at a community college who described having vaso-occlusive crises about five to six times per year. He explained not wanting to discuss his sickle cell disease condition with peers or classmates. He reported having more pain crises when younger.

Dayday is a 30-year-old registered nurse with sickle cell disease who reported having had multiple complications from SCD vaso-occlusive crises. She affirmed having had a stroke, three Transient Ischemic Attacks (TIAs), avascular necrosis, and a hip replacement. She claimed to have had few friends who passed away from SCD. She reported being admitted almost every month for a sickle cell crisis last year, ranging between three days and one month of admission. She reported being admitted every month in 2017 and 2018.

Denise is a 25-year-old lady with sickle cell disease who described the disease as being tiring and frustrating. She reported having frequent VOC, and not being able to do things that other people do. She expressed frustration over multiple and uncountable crises that limit her from a lot of activities.

Jane is a 20-year-old student at a community college. She reported having one to two major VOC and few minor pain crises per year. She explained that her biggest fear is having a pain crisis especially because it comes suddenly without warnings. She stated that taking Hydroxyurea had helped her to minimize the frequency of VOC.

Jaysmoove is a 21-year-old man with sickle cell disease who started experiencing vaso-occlusive crises since his elementary school years. He is a student at a local university, majoring in Multi-Media Study. Mother is a medical doctor while the father is

a businessperson who used to be a professional who played in the National Football League (NFL). He reported having VOC one to two times per year.

Jaz is a 57-year-old lady who has been living with sickle cell disease for years. She reported having multiple VOC per year. She explained that despite the frequency of her pain crises, she would rather stay home and not go to the emergency room because of the negative behaviors of ED healthcare workers. She claimed getting pain relief from holistic remedies. She claimed to have visited the ED only two times in the last year.

JohnLong is a 22-year-old man, a nursing student with SCD who reported feeling normal, but sometimes feeling overwhelmed and depressed related to his SCD status. He feels limited by the disease because he cannot maintain a job because of VOC. He reported having crises about five to seven times per year.

John Martin is a 27-year-old business owner with SCD who makes shirts and sweaters for sickle cell individuals. He declared that with SCD, every day is a struggle because he is constantly tired. He feels people judge him based on his frequent VOC, not knowing the seriousness of what he goes through on a daily basis with the disease. He reported having VOC about 40 to 60 times in a year.

Keisha is a 33-year-old mother of two daughters, with SCD who said she had learned to live with the disease and accept it since the disease would never go away. She also declared that the disease could be annoying and unpredictable. She reported having between nine to ten VOC per year.

KingJeff is a 36-year-old married man, father of three, with sickle cell disease who reported being seriously limited and restricted by the disease. He reported having multiple VOC per year. He reported just being discharged from the ER for a VOC few

hours before his interview. He reported not being able to maintain a job or take care of his immediate family responsibilities as a result of multiple pain crises.

KO is a 30-year-old man, father of three children, with sickle cell disease who experiences VOC at least five to six times per year. He reported feeling depressed because of his limitations related to his SCD and at the same time feeling like a better person because of the disease.

LadyBug is a 30-year-old mother of two children, a boy and a girl, with SCD. She personified the disease as a very tough and rude sickness that has no manners at all. She pointed out having VOC at least nine to ten times per year. She believes that her frequent VOC might be stress related.

Lawa is a 27-year-mother of a newborn, with sickle cell disease who reported having VOC just about 1-2 times per year. She attributed this infrequent VOC to her diet and the way she cares for herself on a daily basis. Despite her SCD, Lawa has a master's degree and owns a business.

Madden Girl is a 24-year-old lady with SCD who reported feeling restricted and discouraged because of her sickle cell disease. She considers SCD as a conflicting illness that can make one feel okay now, and the next minute the person is ill. She reported having at least six or seven VOC per year which sometimes could be greater in number depending on other factors.

Renneth is a 37-year-old occupational therapist who works at a hospital in Broward County. She reported having crises six to seven per year. She explained how vaso-occlusive crises had affected all aspects of her life. She described how hydroxyurea has helped her to significantly reduce the frequency of her VOC.

Tammy is a 32-year-old lady with SCD who just experienced a severe VOC which led to a hospital admission. She declared that SCD had restricted her in so many ways including the area of employment. She asserted that there would be no number to quantify the kind of pain she experiences during VOC.

Shelly is a 34-year-old lady who reported having crises at least five to six times per year. She also reported spending extra years in college because of VOC. She is a professional who works for the port authority. She explained experiencing negative attitudes of healthcare workers in a specific South Florida hospital. She explained that she preferred another South Florida hospital system as she believed she gets better care there.

Warrior is a 25-year-old lady with SCD who considers having the disease as a big responsibility. She also describes SCD as depressing or very stressful as she has to deal with it on a daily basis because it does not go away. She estimated having VOC about 30 to 40 times per year which she considers physically and emotionally draining.

Themes

Transcripts were analyzed according to the systematic procedures explicated by Moustakas (1994). The researcher utilized the epoche process by putting aside any biases and preconceived notions about the phenomenon in order to gain a fresh perspective and see the experience through the participants' eyes. Each participant's experience was reduced to significant statements or quotations and then clustered into themes. Individual textural and structural descriptions were developed, and a composite of textural and structural descriptions was combined to reveal the overall essence of the lived experience of adults with sickle cell disease who have experienced vaso-occlusive crises.

In qualitative research, data analysis involves preparing and organizing the data, then reducing the data into themes (Creswell & Poth, 2017). Themes refer to categories or broad units of data that form a common idea; segments of the data are called subthemes (Creswell & Poth, 2017). The following themes emerged from the analysis that conveyed the experience of adults with sickle cell disease: *Excruciating Pain; Feeling Helpless; Depressing; Dreading Healthcare Workers' Attitude*. (See Figure 4)

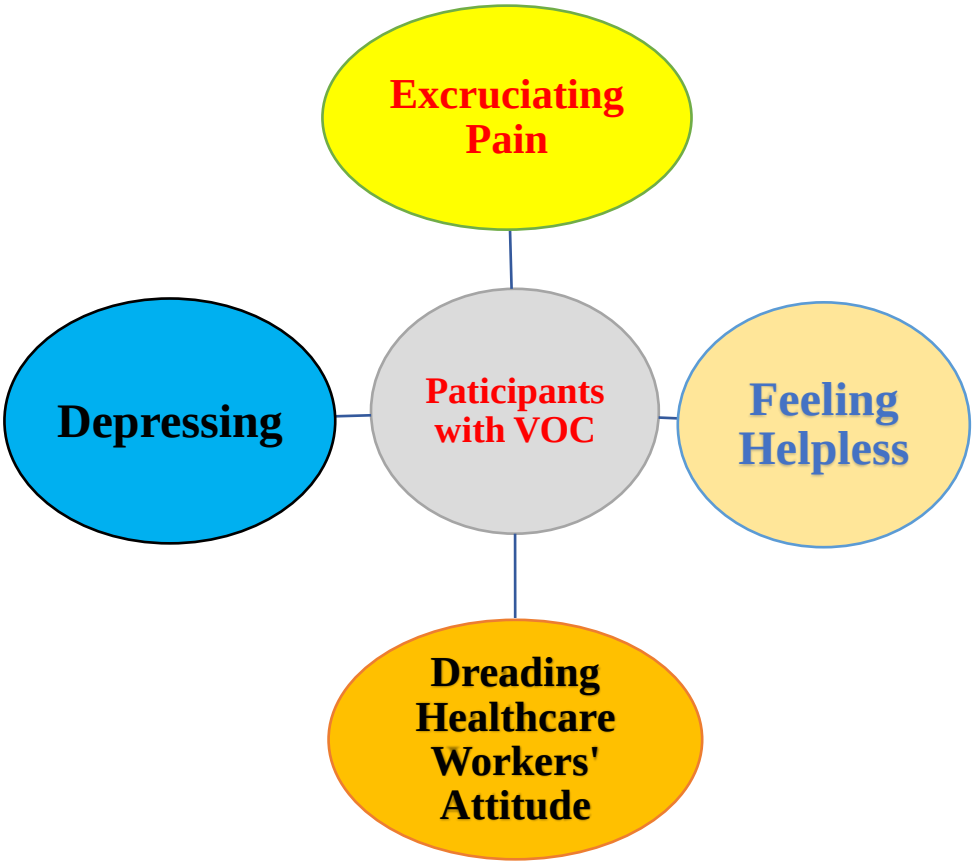


Figure 4. Conceptual presentation of the themes of the lived experience of adults with sickle cell disease (Adeagbo, 2019).

Theme: Excruciating Pain

Excruciating Pain can be described as having to endure an unfathomable and incomprehensible type of pain that can dominate people's lives, and those of their families, friends, and caregivers (Rooij, 2018). According to Lakkakula, Radharani, Henu, and Saikrishna Lakkakula (2018), periods of severe sickle cell pain can last for hours to days, and are challenging to treat and manage, frequently resulting in drastically reduced quality of life. In the following extracts, participants described having to deal with severe, extremely *excruciating pain*.

Shelly described this experience as being very unbearable and depressive. She further described:

> Depending on where you're having the effect of your body could be the severity of the pain as far as from your chest, to your back, to your legs, to your arms, the paining. You feel so much pain, it's like you can't even walk. It's a pain that's indescribable to anybody. A lot of times I can get pains in my chest, it goes to my back and it's just so painful that it's hard to truly explain to people. Because words can't describe it. It is very painful and it's hard to truly explain to people. When they ask you, "On a scale of one to 10, what's your pain?" You can't even say 10. It's past 10, because that pain is so excruciating and to make them understand is so hard because it's just that painful. That type of pain, very excruciating and it's hard to put into words.

Renneth's description of *Excruciating Pain* is reflected in the following narrative:

> There are times where it feels like there's a giant that's 20 feet tall with a giant sledgehammer that's just pounding you in the back over and over. So sometimes it can be like that or there are other times when I'm walking and the pain hits me so bad and so rapidly that it's like I get stuck in the middle of walking either at my house, or somewhere else. And I have to have my husband come and rescue me.

DayDay described *Excruciating Pain* as unbearable. She further described the pain as:

> It feels like "boom, boom, boom, boom", a little like a heartbeat. You're feeling your heart beating wherever the pain is. I also have once in a while, pain in my arm like on crisis, which to me is the most painful. I cannot take crisis. It just more painful because of the cells trying to get through tinier vessels which to me hurt more. So that's how I think that pain is more of a pounding sensation to me, like a like a stabbing, pounding sensation, if that makes any type of sense. I've heard it described different ways, but mine is usually a stabbing, pounding type of type of pain, and it also depends on what my hemoglobin is.

Chris said it is indescribable. He stated:

> I would say it's kind of a weird pain to pinpoint, because most of the time it starts in your back, but you can't exactly pinpoint where it's coming from. It gets worse and worse and then it kind of feels like it's sporadic in different sections of your back. And then when it gets really bad, you can feel it in your whole body and it almost feels like your bones are weak or stiff. It's really hard to describe. It's like a really big ache but it's not coming from one area. You kind of feel in one spot and

> then you feel it on another spot. I'm tossing and turning like when it happens, because I can't sit still.

Jaz declared that VOC pain is the most serious pain she has had to ever deal with in life. She further described the experience as: “As far as being in a crisis, that's a horrible experience. I feel as though I'm lying on a street, a concrete street, and someone is slamming me with an 18-wheeler. That's how it feels, like someone's just slamming this big heavy truck on me. Everything is bone crushing almost”.

John Martin says it’s very intense. He expressed the following:

> Your body just aching. Just, it's like sometimes the pain could be intense, like a sharp pain and you probably couldn’t deal with it, it's not steady sharp pain just like a little sharp pain here and there and you deal with it. But some of it, it's like this stabbing, like somebody's got a knife and stuck it on your skin and chomping on top of it. And it's not one person doing it. It's like they have a line up and people, one person get off of it, then other person is already in the air and about to land on it and it just like is constantly.

Warrior described *Excruciating Pain* as a big responsibility, stressful, and depressing. She said:

> The pain itself is like someone just repeatedly hitting you over and over again. Maybe with a metal bat or, even if you was in the car accident, it's just some pain that you cannot get away. It's just throbbing, aching, and sharp, and it's just back to back. Back to back. It's worse than a migraine because I know a migraine, you had been going back to back, back to back. But it's way worse than that because,

> instead of just your head, it's from your head to your feet, so it's like your whole body.

According to **Keisha,** VOC pain feels severely and extremely excruciating. She described it as:

> As I get older, I realize now it feels more like I'm about to have a heart attack when I have these pains. It's so bad that I feel like my heart is about to stop. But, it's just horrible. It's like you feel like a bunch of people stabbing you. Somebody's stabbing you, and then you start to feel like your joints are locking up. It hurts so bad, but it feels like just somebody's just stabbing you nonstop, just a bunch of people stabbing you in different locations of your body. You fall asleep, and then, you wake up, and it's like somebody just wakes you up stabbing you again.

Madden Girl described *Excruciating Pain* as:

> Usually I feel like I am getting ran over by an 18-wheeler truck. It's just going forward and backwards on me. It's not easy to describe. You feel like somebody has taken a knife and it's just repeatedly stabbing you. Every area of your body, no area is off limit. Not your buttocks, not your toes. Everything is affected. You feel like your bones are snapping. It's like somebody is just crushing you. Sometimes your chest feels so heavy, you don't even have to take in air.

Brad described *Excruciating Pain* as a sudden pain that originates from the back and makes one moan and shake. He further described:

> You sit down and you want to get back up, but you can't get back up or you just have to find a sweet spot within the pain. You're going to feel pain through the whole thing, but whatever spot you can sit in that makes it feel like less pain is what spot you have to find to make yourself feel better.

The sickle cell patients in this study verbalized *Excruciating Pain* during vaso-occlusive crises. They explained how agonizing and unbearable the pain of VOC is experienced. They all highlighted the type and severity they experience during VOC, and their coping mechanisms.

Theme: Depressing

Depression can be defined as feeling of sadness or low mood, loss of pleasure in or interest in daily activities, and reduced energy nearly every day; together with having some symptoms as: feelings of guilt, reduced self-esteem, disturbed sleep, problems in concentration, change in appetite, change in activity, impaired function, ideation of self-harm (Jokelainen et al., 2019). Simsek, Evli, and Uzdil (2019) explained that pain plays an adverse role in the psychological status of the individual suffering from it. They further indicated that cases of chronic pain such as that experience in VOC are most frequently accompanied by such symptoms as depression, anxiety, anger, helplessness, aggression, and despair all of which causes the individual to be isolating.

Tammy expressed wanting to give up during vaso-occlusive crises because of the intensity of her pain. She described *Depressing* as:

> During pain crisis it feels like I want to die. I'm stressed, I'm depressed because nobody in the house understands the way I feel. So I just want to crawl and die

> because the pain is so much for you to bear. You don't want to live. It's painful and people always think, "Oh yeah you're depressed." Yes, I'm depressed. Nobody could relate to me in the household. They cannot comfort me. So yeah during pain crisis I do want to die. So that's how majority times I feel, when I'm in crisis. I'm alone so obviously.

JohnLong described his mental status during VOC as below:

> Mentally, I feel super depressed because sometimes I have to get things done and I have to postpone, or I can't do it because I'm in crisis. The pain, sometimes it'd be unbearable, sometimes I can manage it. It just makes me feel terrible sometimes.

KingJeff expressed *Depressing* during VOC. He feels alone when no one is there to help. He states:

> Because when you don't have nobody to talk or you don't have nobody there you don't have a support system, it's like you're all alone. You feel like giving up. It's like the only thing you want to do, is just lay here and die. Because you don't have nobody to talk to. If you find that you feel like nobody care about you, you feel like you've been abandoned, you are a lost child.

Renneth explained that she gets sad during crises because she feels she is putting a lot of burden on her spouse. She expressed feeling isolated during crises. She explained:

> But I feel a bit sad too and a little bit on the depressed side also during those moments too because then it feels, it's like I'm putting a lot on my husband because he has to deal a lot of the work around the house and I felt bad because

he's already doing a lot in my house anyway. It is sometimes lonely during crises, so I have a time or two I had gotten out of bed and tried to get to the bathroom, The other time it was not so lucky and then I totally peed all over myself because I really had to go and there was no one to help. So it's just nice to get a little attention every once in a while. I'm a very patient person so it's not like you have to be on top of me every two seconds, but every half hour or so, just be like, "Okay, do you need to use a bathroom? Is there anything that you need?" Just to kind of, you know, just say hello, see how I'm doing. That's always nice.

Warrior explained that VOC is depressing and isolating. She explained:

My last few visits to any hospital that I went to is I literally have to sit here in the ER, never been triaged, and have to wait four or five hours. Then, you finally get to the back and the doctor take another hour too. To me, that's just too much just to be sitting there all in pain and they're not doing nothing. You feel depressed, you feel lonely, you feel isolated. Then, when you get to the back, the doctor thinks you just there for meds, so you have to wait for your, you have to wait for your blood results to come back for them to take you serious that you're in a crisis. I was like, I just avoid ERs. Very depressing.

LadyBug described having VOC as depressing. She states:

There are times where, you really sit and think that it's hard to be in pain 24/7, and you're not with your kids as you should be. In the hospital and away from them. It's kind of depressing a little bit. You can't really let it get the best of you. You got to keep going, that's why I keep a picture of my kids as my screensaver

> because they keep me going. Or it will just be stress, and I'm trying to balance everything all at once with no help and that'll really send me back. Meantime, I'm trying to learn how to control my stress level. That's my biggest downfall that I'm still trying to get control after all these years. So in the meantime I am looking for a part time job, so I can have a little bit more income. Because you know, the disability that can only, you know, cover oh so much in 30 days. Yeah. I can't cover everything, they cover the majority of the bills. It's really hard and depressing, but I'm getting the hang of it slowly. get chosen to deal with this?" Then again, I remember when they say, "God wouldn't give you more than he know you can handle."

All participants shared their emotional and mental feelings during VOC. Each one of them explained how depressing and isolating vaso-occlusive crises could be, as they are the only ones experiencing the pain of VOC.

Theme: Feeling Helpless

According to Madubata, Odafe, Talavera, Hong, and Walker (2018), helplessness is one type of cognitive appraisal that can emerge when an individual perceives certain events as uncontrollable. They further explained that helplessness is a coping response to stressful situations and results in negative emotions and maladaptive behaviors. It is a cognitive response to repeated exposure to stressful and uncontrollable events. Individuals who develop learned helplessness experience negative motivational, cognitive, and emotional effects. The researchers asserted that there is an association between helplessness and depression, suggesting that learned helplessness is a result of cognitive exhaustion due to exposure to uncontrollable situations (p. 626).

Renneth expressed *Feeling Helpless* as a result of pain VOC. She explained how her husband had to help her during pain crises:

> I kind of feel like not small, but helpless. The other time I was not so lucky and then I totally peed all over myself because I really had to go. So it's just nice to get a little attention every once in a while. So sometimes it can be like that or there are other times when I'm walking and the pain hits me so bad and so rapidly that it's like I get stuck in the middle of walking either at my house, or somewhere else. And I have to have my husband and come and rescue me. I'm basically stuck to a bed, which I don't like being. It's like I'm putting a lot on my husband because he has to deal a lot of the work around the house and I feel bad because he's already doing a lot in my house anyway. But I also need someone to be compassionate and actually listen to what I have to say and not just write me off just because of whatever. I need someone to be compassionate and actually listen to what's going on and see what I may need and try to think of different ways to help.

Shelly explained that during a crisis:

> You feel helpless and sometimes you feel really defeated. The few times I've been in the hospital, I more or less felt more of a helpless, depressive spirit because of that simple fact that is like, I don't want anyone worrying about me. I'd rather try to be this strong person and act like I can bear through it, but sometimes you really can't. That makes you truly feel helpless, and I'm here, I'm depending on everyone.

Cerena explained that she needs family and or friends when she is in crises. She further explained:

> Definitely need someone that's going to be kind and patient and understanding. Feeling the comfort of whoever's there. It doesn't even matter. There are times where I've gone on school trips and gotten sick and it was the people that were around me. My advisors or teachers or professors whoever that were there, and they were kind and comforting. I perceive more kindness from strangers in certain cases. It doesn't really matter. It's just that kind of mitigates the loneliness and isolation that you feel when the crises start to come on, just having someone there, like, "Okay, do you need water? Do you need this? What can I do?" Just the question alone, just having friends that are like that too, it's really helpful.

John Martin voiced:

> Somebody that's there to support you, you need that. You need, always want somebody, one person there, they'll support you. Somebody that's there to help. If you can't do nothing, you can't talk, somebody to help that you already done explaining what's going on with you.

JohnLong verbalized: “it's like sometimes I want my family support to be there to support me and tell me things are going to be okay.”

Dayday explained her *Feeling Helpless* as:

> But when it comes to like more of a psychological aspect, it's just have been my immediate family, which is really just my parents and my grandmother. They're what I call my food runners, my back rubbers, my, "Can you go home and get my glasses so I can take out my contacts?" So it's just having someone there to kind

> of keep you sane, keep you comfortable, help you do like the little things. So it's just having someone there to kind of keep you sane, keep you comfortable, help you do like the little things. So that's like the main care that I really need, like more from family supports.

Lawa described *Feeling Helpless* as:

> The pain is something like, I feel like I'm in another state of mind. Like I'm not even here because I'm just down. I'm always in pain. So I'm not always in the right mind. I'm always down. The pain is something I never experienced before. It's very painful. I will always say that emotionally, I'm always down. I'm never happy. I'm always crying. I always have an attitude. I don't want anybody talking to me. The most person I probably would want around is my dad, because I'm a daddy's girl. But my mom was the one who is always usually doing everything for me when I'm sick because I can't help myself. But for some reason, the sickle cell pain always makes me have an attitude towards people. I don't know why, it's not good.

Most participants in this study expressed feelings of helplessness during a VOC. They also spoke of needing support from family and or friends, claiming this sometimes uplifts their spirits. They explained how important it is to have someone around during vaso-occlusive crises.

Theme: Dreading Healthcare Workers' Attitude

Attitude is defined as a behavior based on conscious or unconscious mental views formed through cumulative experience and a complex mental state involving beliefs,

feelings, and dispositions to act in certain ways (Altmann, 2010). According to Jenerette, Pierre-Louis, Matthie, and Girardeau (2015), People with SCD have reported being stigmatized when they seek care for pain. Nurses' negative attitudes is one of the factors that contribute to stigmatization, and may affect patients' response to treatment, care-seeking, and eventually patient outcomes. Bergman and Diamond (2013) argued that the undertreatment of pain during VOC is often associated with health care disparity issues and that pain complaints of racial and ethnic minorities are less likely to receive adequate attention. Suspicion and perceived drug dependence combine to negatively influence sickle cell patient care in the clinical setting. The beliefs and attitude of ED medical professionals are critical in determining the quality of care that a SCD patient receives. The participants shared their experience *Dreading Healthcare Workers' Attitude.*

Dayday spoke about her experience with healthcare workers and their attitudes during some of her crises. She expressed *Dreading Healthcare Workers' Attitude* as:

> I get nervous going to the hospitals because I don't want to go through the stress of going through emergency room with the ER doctors. They tend to always view you as a drug seeker no matter what. I'm allergic to morphine. So once I say I'm allergic to morphine, they automatically think "Drug seeker. She wants dilaudid", and it goes from there. The care, to me in the emergency room is more based on a stigma or stereotype of sicklers being drug seekers or frequent flyers. It's another terminology they like to use.

King Jeff shared his experience with some healthcare workers during crises. He described:

> A certain doctor or certain doctors, when I check in, by the time they see me, they're like, "Oh no, I'm not... I can't give you no narcotics and I'm doing nothing for it. I could draw your blood to see if you're sickling really bad." It's always the people who you run into at the time of the visit, because some doctors look at you and they're not going treat you. All they're going to do is draw blood and send you on your way. You'll be sitting there for two hours waiting for your blood results come back, in very bad pain. I walk out on them. I'm not going to sit in no hospital waiting two, three hours just to see what my pain level is.

KO described how bad he was treated during one of his vaso-occlusive crises. He described:

> A couple of days ago I was at Broward general. The doctor came and ran my blood and he was like 'um, your blood is fine, you can go, you don't have sickle cell'. I said, so are you telling me I don't have sickle cell because my blood not low? He said yeah, there's nothing wrong with you, your discharge paper is coming up. So I'm thinking to myself like wow, just because my blood's okay, you telling me I'm not in pain? That don't make no sense. So, it really depends, some doctors are very nasty, they are very nasty. And it's a shame, it really is a shame. They really don't try to help us.

Renneth expressed her frustration with healthcare workers during her crises. She explained *Dreading Healthcare Workers' Attitude*:

> The one thing that you have to deal with is some of the nurses being like, "Oh, she's only there for of pain medication. She's a druggy. She's addicted to the

morphine or she's addicted to some kind of pain medication." And having to deal with, "Okay, the last time I was in the hospital was how many months ago or a year ago or whatever. If I was really addicted to pain medication, wouldn't I be in here more often?" Yeah, you are going to have some that are addicted to it and just fake a crisis just to get the pain medication. But most of us don't.

Jaysmoove explained his experience with attitudes of some healthcare workers:

The staff members are sometimes rude too. Sometimes they can be a little overwhelming. Sometimes they can also ... They don't understand where you're coming from. It's hard for somebody to deal with and treat you a certain way you expect when they've never actually been through what you've gone through. There'll be extreme pain that I'm experiencing. But maybe sometimes the care wouldn't come fast enough or wouldn't come as convenient as I expected it to be.

Lawa voiced her concern regarding *Dreading Healthcare Workers' Attitude*. She said:

And I went to adult starting at the age of 22. From a children hospital's department and going into an adult hospital, it's a different ballgame. I can't say the same about adult. Some of them think I'm a drug addict. They don't want to give you pain medicine, or if you tell them that you're in pain, they don't believe you. Like I mentioned, actually that happened about twice when they kept me in the ER for 24 hours. I'll speak more of the doctors, the nurses, the LPNs, CNAs. Sometimes I don't want them around because I remember this one incident. I wasn't feeling well, and the nurse was just giving me attitude the whole time and I was asking for a new nurse and they wouldn't give me a

new nurse. I remember I was in the ER for 24 hours, no medicine, no IV fluids, nothing. They just put me in one corner.

All participants expressed their concerns, worries, and fears of negative attitudes from healthcare workers. Most of them spoke about their fear of stereotyping and stigmatization from healthcare workers, especially in the emergency room. Most participants are still *Dreading Healthcare Workers' Attitude.*

Individual Textural and Structural Descriptions

Using Moustakas' (1994) transcendental phenomenological approach, the essence of the phenomena is derived from the participants' experiences. The researcher's role is to generate the textural and structural narratives for each participant. The textural description explains "what" was experienced and the structural description describes "how" the experience occurred (Moustakas, 1994). The textural and structural descriptions of each participant are then synthesized into a composite account of the experience. The final step in the phenomenological process includes integrating the textural and structural descriptions into unified statements of the essence of the experience (Moustakas, 1994).

Brad's Textural Description

Brad is a young man with SCD whose twin sister also has SCD. He described his experience having SCD as:

> I mean I personally don't have any particular feelings toward having sickle cell disease because I just wake up and don't remember that I have it to be honest with you. It's only until you feel pain or there's a certain limitation to the things that

> you do that other people are doing that you realize that, "Oh right, I do have that disease. I do have sickle cell." Until you actually have to go through something, you're going through a crisis, you're experiencing a symptom. So, a lot of times people don't even notice that I have everything wrong with me until I have a crisis. That's when I realized that's what comes with having sickle cell.

Brad's Structural Descriptions

Brad described having significant crises only seldom. He elucidates that when he actually had significant ones, they were always very severe. He explained his experience during crises as below:

> I just remember the symptoms, but sometimes I'll feel an ache that is a slight thing, but when the real bad ones happen, like every few years, I want to say it was like around December, January time, I was in the hospital for the longest I could remember. They were trying to help me feel better again and things were helping, but a lot of things weren't helping. Because what happens is like the pain makes everything around that area sore and it had started in my back, that's where it started, so your hips, like you can't really flex them how you used to. So, I remember I couldn't walk after, that's what I remember, and I had to learn how to walk and it was just a terrible experience, I have a heating pad in order for it. But mentally, it's really draining.

Cerena's Textural Descriptions

Cerena asserted that SCD has affected her life in so many ways. She believes everything else in her life comes second because SCD comes first; SCD comes before everything else. She further described her experience with SCD as below:

> Like I said, everything else comes second to sickle cell. I don't even know and now that I am able to manage a little bit better and I'm actually able to make a budget and stick to it and call my friends and go out and do things, I'm like, I don't even know how I made it through my first 30 years of life. My money is a mess. My life is a mess, because as soon as I get sick, which is constant, just back to back, you don't have time to process anything else. I have to deal with this so I can turn in my assignment a little bit later, or my mom is going through something. I can sit and talk with her and comfort her. Everything else comes second to sickle cell. The last time I had a job, they fired me while I was in the hospital.

Cerena's Structural Descriptions

Cerena explained that she used to visit the emergency department for VOC pain multiple times every month before she started taking different holistic supplements. She declared it's been almost a year since she had been hospitalized for VOC because of the alternative therapies that she uses. She further described her experience having VOC as:

> During crises, everything in life goes on hold. It doesn't matter what's external, what's happening in the outside world. Everything just kind of stops and comes secondary to the pain and to the issues, so I've had necrosis of the hips, so the blood flow to the hip bone dies. The connection dies, and it's very severe and it hurts terribly. That's one of the most painful ones that I've experienced, painful illnesses and issues that come up as a relation to sickle cell, but in general, usually I experience back pain, joint pain.

Chris' Textural Description

Chris is a young man with SCD who would rather not talk about his condition because of fear of stigmatization. He described his experience having SCD as:

> It does depend on how severe your sickle cell is, because there are people that have it worse than some people that have it, their sickle cell is not as bad. I could say there's probably times where I thought I was going to die when I was younger, and I got really sick. So when you're younger, I guess you kind of have that in the back of your head. Physically, I would say you feel the same except for when you do have the crisis. Aside from that, I would say you probably feel like the average person.

Chris's Structural Descriptions

Chris reported having crises multiple times in a year. He claimed that his pain crises were more frequent when he was younger. He explained his experience during VOC as below:

> I would say it's kind of a weird pain to pinpoint, because most of the time it starts in your back, but you can't exactly pinpoint where it's coming from. How do I describe this? I wouldn't say sharp, but it kind of feels like a backache at first. But it gets worse and worse and then it kind of feels like it's sporadic in different sections of your back. And then when it gets really bad, you can feel it in your whole body and it almost feels like your bones are weak or stiff, when it gets really bad, I would say. It makes you feel like you can't sit still. It's so sporadic though. I would say it's kind of hard to explain. It's really hard to describe. It's like

> a really big ache but it's not coming from one area. You kind of feel in one spot and then you feel it on another spot. I'm tossing and turning like when it happens, because I can't sit still. Yeah, definitely can't sit still when it happens.

Dayday's Textural Descriptions

Dayday affirmed that SCD has pros and cons for her because if it were not for SCD, she was not going to be a registered nurse today. She reported SCD as totally exhausting. She explained having lost few friends to SCD. She described her experience with sickle cell as:

> Well for me there's pros and cons in the sense of, I feel like if I didn't have sickle cell, it wouldn't make me the person I am today. I'm 30 years old now and I've already had one stroke, three TIA's, have a vascular necrosis and I've had a right hip replacement. So I've had a lot of physical complications that came along with the disease. I've lost many friends from sickle cell. My best friend from high school had sickle cell also. She had a stroke and so passed away, so I've lost many friends also do sickle cell. So in so in a sense, it's just really hard, and it's a very complicated disease. I'd be like is not really understood by healthcare professionals and that's what made me become a healthcare professional myself. I realized how healthcare workers, look at sickle cell patients, it is as if they feel as if the disease is supposed to disappear. So always felt like maybe if I continued my education and furthered myself, I show that I have this chronic disease, but I don't let it stop me. Physically, it's just draining, in a sense of I just want one day that we don't feel any type of pain or the complications that also come along with the disease.

Dayday's Structural Descriptions

Dayday described having VOC almost every month especially in 2017 and 2018. She explained that most of her crises lasted between three to four weeks. She said the frequency has slightly reduced this year. She further described her experience during VOC as below:

> And so my pain is probably like at a so-called like maybe fifteen or something on a pain scale. So, so far this year I've been to the emergency room roughly three to four times, and I'd been admitted each time I went to the emergency room. Usually if I goes to the emergency room, I'm admitted, nine times out of ten I am being admitted. Sometimes I choose to, I would just take like a few doses of pain medicine to where I feel comfortable with my pain and I can go back to my oral pain medicine cause I don't want to be admitted in that sense.

Denise's Textural Descriptions

Denise is a young lady who explained that SCD has restricted her in various ways. She reported not being able to do many things that she likes do because of the disease. She further explained her experience being a SCD patient as below:

> It makes me depressed. Being sick all the time, not being able to do things, missing out on stuff. So those things, it makes me sad. It's depressing, it's frustrating, because I missed out on a lot. I really missed out on a lot. Financially, I know for a fact, if I go get a job I'm not gonna stay while I get sick too often. So I get government assistance, so that's what I'm living on. Without that, I don't know how I will manage.

Denise' Structural Descriptions

Denise reported having multiple VOC per year. She reported having pain crises almost every day. She further explained her experience with VOC as:

> Most mornings, I wake up in pain. Almost every morning, I wake up in pain, but it's not always bad. It's the bad pain that I have to go to the ER for, but generally with sickle cell, you're going to have pain most of the time, but certain pain, I'm able to deal with it, I'm able to cope, I'll go take a hot shower and things like that. And then you have the bad pain, the real crisis that you have to go. You have to take something, like pain medicine, and then you have the real back pain, like pain medicine's not helping you, warm water not helping, nothing helping, so you have to go to the hospital. To be honest, I can't even count how much times. You breathe wrong, it's hurting you.

Jane's Textural Description

Jane is a young lady with SCD who has a twin brother who also has SCD. She and her twin brother realized they both had SCD at about five years old. She described her experience having SCD as below:

> It doesn't feel any different, I would say. It only feels different when I have a crisis. So I live a normal life, there's nothing that I really can't do. I have to take extra precautions sometimes; I have to drink more water. I take my medicine every day. But other than that, oh and I also have to make sure I don't overexert myself. But other than that, I pretty much live a normal life, like average. And yeah, that's pretty much what it's like.

Jane's Structural Descriptions

Jane reported having crises less than one time in a year. She reported her last crisis was in May of 2018. She believes that the infrequency crises is probably because she takes hydroxyurea. She explained her experience during crises as:

> So I'll just tell you about my last one. The last one that I had; I'll just describe the feeling. It was like tightness in my chest. It was more like my rib cage, chest area and it wasn't necessarily pain. There was different moments where I would feel shooting pain somewhere in my body. But for the most part it was just tightness in my chest and sitting up or standing up was difficult because you use your midsection to do those kinds of things. So it was difficult to even breathe, to take deep breaths. I couldn't take deep breaths because the pressure was just so intense.
>
> The pain started in my lower back and I did take Ibuprofen and so I tried to go to sleep. I remember laying on my stomach I think. And then I felt pain shooting up to my neck. So I actually change my position and it wasn't working. So I just sat in a chair and then that's when the pain in my legs started. And so I was just tossing and turning. I fell asleep a couple times for like maybe an hour or 45 minutes, but I'd be up again. And then so yeah, that just kept going.

Jasmoove's Textural Descriptions

Jasmoove was told by his parent that he had SCD when he was in elementary school. He explained his experience having SCD as:

> Well, having sickle cell disease, for me it's a thing, I mean, obviously, I feel more limited. So I would say even as far as like stamina endurance, I am limited. As far

> as stamina, a normal person might have, on a scale of 1 to 10, their maximum stamina could be 10. I feel like with me, my maximum could be more so, even a seven. I have a shorter maximum than most people just because I have sickle cell. Sometimes there are even times where I don't feel as energized as I should. Sometimes I feel kind of drowsy when I wake up.

Jasmoove's Structural Descriptions

Jasmoove is a SCD person who reported having sickle crises about one or two times, sometimes three times per year. He described his experience during VOC as:

> Sometimes, if it gets really bad, I feel kind of helpless. There's nothing that can even help me. I do feel sometimes, I feel like I need hydration. Every time I do have a pain crisis, I feel like I just need to hydrate myself and just drink water. It's definitely a lot of aches. I would say on a scale of one to ten, ten, if it gets really bad. For the priapism, they told me if it prolonged over two hours, go to the doctor. If it doesn't go for long than two hours, there are definitely a lot of aches. It feels like just pain against my will, like pain that I didn't cause, it's just unintentional. I don't know. I definitely feel a lot of aches. I feel a lot of tension in those parts of my body that are affected.

Jaz's Textural Descriptions

Jaz is a 57-year old lady with SCD who declared having had difficult times with SCD especially during college years and during her only pregnancy period. She pointed out that she had come close to death few times because of the disease. She further described her experience living with SCD as:

> College time was kind of interesting. Stressful, of course, and kept me in the hospital all the time during college. My pregnancy, that was horrible. I spent the last three months on bedrest in the hospital, crises all the time. As soon as he started getting bigger and the blood exchange, forget about it. Always sick, completely, always sick. So during a surgery, I had a major heart attack, okay. The crisis is, it was horrible. The priest came up, read me my rights. They thought I was going to die. I'm telling you, I had people all around there. The priest had his Bible, the holy water, and he's reading me my last rights. They really thought that I wasn't going to make it.

Jaz's Structural Descriptions

Jaz admitted having VOC as many as nine to ten times a year, but having visited the ED only couple of times in the past year. She explained feeling short of breath during vaso-occlusive crises. She further explained her experience during VOC as:

> During crises, can't breathe, first of all. I feel like my lungs, my chest feel compromised. My head feels compromised. I've had MRIs. I've had over 35 small strokes on my brain that shows up now on MRIs and stuff, so when I go into a crisis, immediately my head starts feeling... It's not even like it hurts. It feels sore, like someone's kind of like... not like a, you know you have a headache. It's almost like if it's compressed like in a small bottle. It feels like pressure.

JohnLong's Textural Descriptions

JohnLong explained that SCD had limited him in many ways. He feels the disease has limited him significantly in some areas of his life. He affirmed that areas like

employment, education, and finance have been negatively affected by the disease. He described his experience having SCD as below:

> It limits me. Like working, it limits me finding jobs. When I get sick and I have to call off, some job employers will feel like I'm being lazy when I'm not. Sports, I love sports, but I really can't do all types of sports and stuff. It does limit me. You want to do certain things, but because they know what you're going through, they hinder you from doing those things sometimes. I know financially, it affects me a lot because I can't hold a job and when I do get a job, I don't work that long because I'm calling out because I'm in pain or stuff like that. You want to do certain things, but because people know what you're going through, they hinder you from doing those things sometimes.

JohnLong's Structural Descriptions

JohnLong described having VOC about five to seven times a year. He explained being limited by crises and not being able to do many things because of VOC. He further described his experience during VOC as:

> For school, it's an issue sometimes because I have pain before class and then I'll take an opioid pain medicine and now my concentration is messed up, so I'm just there listening but not grasping the information. Certain aspects, it really do affect me bad. Mentally, just sometimes there's depression there, but I don't dwell on it for too long. I just accept the fact that it is what it is. They make you feel weak. Mentally, I feel super depressed because sometimes I have to get things done and I have to postpone, or I can't do it because I'm in crisis. The pain, sometimes it'd

be unbearable, sometimes I can manage it. It just makes me feel terrible sometimes.

John Martin's Textural Descriptions

John Martin is a young man with SCD who explained that SCD has a lot of restrictions on his life. He ended up having to establish a because no employer wanted to accommodate his SCD and the associated crises. He further described his experience as below:

> It feels like some days you just can't, you can't cope, you can't get out of bed, you can't go do regular stuff, you can't really chill with friends. It stopped me from working a lot. Yeah. Sometimes you can't get up to go to work. And a lot of companies don't see that they don't know about it. So you know, sometimes you got to push at work. Sometimes you have to work in pain, the people around you and just regular, normal happy and you the one, you sitting in the back, bones hurting, body hurting, and you're still here and you got to keep pushing sometimes. Sometimes you want to take trips to go out and do stuff. You get to the point where it's time for you to get to that trip and you're in pain. You can't go. Sometimes you're stuck in the house watching movies, dealing with a little pain because you can't do nothing.

John Martin's Structural Descriptions

John Martin described having VOC pain at least 40 to 60 times per year. He described his experience during pain crises as below:

> I could wake up and have a small crisis in my elbow and my arm or something. Something I can deal with on my daily basis and just, deal with it and cope. But two days from now I have a big crisis, somewhere else in my body and go to the hospital and they treat that. But then two days later from that, I have another small crisis that I just have to deal with. And it's just, it's an everyday battle. You don't know what's coming to you every day. So it's more than 60 times.

Keisha's Textural Descriptions

Keisha declared that she had learned to live with SCD and accept the fact that she has to live with the disease. She also said that the disease could be very annoying. She further described her experience living with SCD as:

> I've learned to live with it and accept it. That's one thing. That's the first thing dealing with sickle cell. You have to accept the fact I have this, and it's not going go away until I'm dead. Secondly, it has its annoying moments when you least expect it or when you're just fine right now. You're bouncing around. Next thing you know, you're balled up on the floor in excruciating pain, and it's like, oh my gosh, not right now. I've found a way to cope with it. I try to bounce around, and I try to live the most happiest life with it.

Keisha's Structural Descriptions

Keisha declared having more than ten VOC in a year. She explained that she is able to deal with small crises and take care of them before they become big. She further described her experience during crises as below:

> And then, you get to the point you can't even walk. It's just a lot. But, I just fight mentally, and I'm not going to allow my legs to lock up. I'm not going to allow my legs to lock up. I'm not going to allow my arms to lock up. I'm not going to allow this. I have to walk.

KingJeff's Textural Descriptions

KingJeff is a father of three children who explained that his family did not really understand SCD, and that he had to try to understand the disease by himself when he was younger. He further explained that the disease had limited him in various areas including the areas of employment and family responsibilities. He explained his experience as below:

> Growing up with sickle cell, it was always bad because I didn't really have support system there. My grandmother, she didn't really understand what sickle cell was because she's Haitian and she full blown Haitian. So I had to grow up knowing what sickle cell was for myself. It's affected me really bad because I can't keep a stable job to take care of my family. I can't maintain a good lifestyle because I can't do the sports. I grew up really doing bad because of my sickle cell. I can't interact with my kids the way I want to. I have to be careful with certain things I do or certain areas I go in life.

KingJeff's Structural Descriptions

KingJeff reported having multiple VOC every year. He described having crises at least six times per year and being admitted in the hospital at least two or three times per year. He further described his experience during VOC as:

> I feel terrible. I do, my pain crisis, I feel terrible because when I'm going through my pain crisis, I don't want to interact with nobody. Because the only thing that's on my mind is trying to get my pain better. The pain crisis I go through is very, it's very excruciating. So all my pain attacked me, everything in my body hurts. So, from vascular necrosis to 26 screws in my back and going through sickle cell crisis at the same time, it's very excruciating.

KO's Textural Descriptions

KO described having SCD as depressing and limiting. He reported not being able to do the things he would like to do because of his SCD status. He additionally described his experience having SCD as below:

> Um, like I said it's depressing. Cause there's things like right now, I can't just go out there and just get any job, okay, because of my situation. Sometimes, I go work and then I might have to miss a whole week off of work because I'm in hospital. So, it's depressing because, as a man, I can't provide like I want to for my family. Because of my situation. Um, I mentioned before, if I get a job, I go work at Metro or something, and I have a crisis and I have to go to the hospital, I might be out of work for a week. I mean that's cutting out on my paycheck so for me to provide for my family.

KO's Structural Descriptions

KO explained that his pain crises occurred multiple times per year, both mild and severe crises. He described the pain crises as severe and excruciating. He further described his experience during VOC as:

> Pain wise, it's tiring, it hurts very bad. It feels like every bone in my body is breaking at one time. And it's everywhere. It is depressing. But then again it does make you want to, you know, it does make you feel like a better person. You know because I want to become a better person because this disease, there's only certain things that we're limited to do.

LadyBug's Textural Descriptions

LadyBug explained that living with SCD is sometimes depressing because she had been restricted in so many ways by the disease; she voiced out that sometimes could not be with her children because of VOC. She further described her experience living with SCD as:

> It's basically, you wake up every day with some type of pain, whether it's in your hips, like me or everywhere else. But that's my main pain, every day I wake up, it's my hips that bother me. So psychologically, nothing, just live your life. Some people say having sickle cell doesn't bother them; some people will tell you. There are times where, you really sit and think that it's hard to be in pain 24/7, and you're not with your kids as you should be. You are in the hospital and away from them. It's kind of depressing a little bit. It basically, it gives me limits to what I can do and how much I can push myself, so I don't end up in a crisis.

LadyBug's Structural Descriptions

LadyBug declared having more than ten VOC and ED visits per year. She further described her experience during pain crises as:

> Honestly, it feels like someone's just hitting you over and over with a bat, wherever your pain is. I can say my pain is in my hips and my knees, so it feels like someone's just hitting me in my knees, very annoying pain. The weather, especially when it rains, makes it worse, so my knees tend to give out or my hip tends to give out on me so I would have to hold on to stuff. But I'm saying it's just constant pain, every day.

Lawa's Textural Descriptions

Lawa is a young woman who used to be ashamed related to her SCD status because of how she got picked on by her friends in school for the disease. She attributed her infrequent hospitalization to her healthy style of life. She further described her experience having SCD as below:

> Growing up, I used to think having sickle cell disease was, I used to be ashamed of it. I didn't like it because I used to get picked on when I was in school. But health wise, it's nothing nice, it's nothing fun to have. It's hard, it's very crucial. I know how to handle my health better like the importance of drinking water. I know if it's raining, I have to cover myself up or I can't be in water, or certain things I shouldn't be eating. I stopped eating junk food at the age of probably 13, because I knew it had an effect on my sickle cell. I won't eat out. I won't eat McDonald's, I won't eat Burger King, I won't eat any of that because I knew it had an effect on my health.

Lawa's Structural Descriptions

Lawa reported having VOC only 1-2 times per year. She described her experience during VOC as below:

> The pain is something like, I feel like I'm in another state of mind. Like I'm not even here because I'm just down. I'm always in pain. The pain is so bad you can't even describe it sometimes. But some people be like, "Oh, what is your pain, one through 10 or what does the pain feel like?" And it's just a pain that you can't describe. It hurts so much that you don't even know how to describe it. So I'm not always in the right mind. I'm always down. The pain is something I never experienced before. It's very painful. I will always say that emotionally, I'm always down. I'm never happy. I'm always crying. I always have an attitude. I don't want anybody talking to me.

Madden Girl's Textural Descriptions

Madden Girl expressed having SCD as tough, restricting, discouraging, and conflicting. She believed that SCD is also a gif. as the disease has made her stronger. She further described:

> Yes, discouraged. It's hard to put it in words because there's so much that goes into sickle cell, but I would say in a sense, I feel gifted sometimes. I will say that. It's tough, but it builds you. It helps you become stronger. It allows you to see the outside world differently from the regular person who has nothing going wrong in their life as far as a medical illness goes. It's also tough… It's really tough. Sometimes you feel like you're alone in it, but you're not in a sense because there's so many other people out there that has it, so you kind of have the support. It's about feeling comfortable enough to reach out to talk. It's a conflicting illness. You want people to be there for you, but then at the same time you feel like a burden to people, so you have to figure out how to manage the two. It's tedious,

> but you do it anyways. You have to find that will to say, okay, I have an illness that is not regarded in the media as anything serious, even though it's life threatening. You can be fine today and tomorrow you're in a coma or something, so it is very serious. When you have an illness that restricts a lot in your life, you become more appreciative of those little things that others would overlook.

Madden Girl's Structural Descriptions

Madden Girl described that she has VOC throughout the year. She explained that she has severe VOC at least six to ten times in a year. She described:

> I spent a lot of time fighting myself, fighting my body, like, this shouldn't be happening to me. Why is this happening to me? And I had to come to terms with understanding this is going to happen. This is what I have. This is what it is. I need to adjust my mentality. I need to adjust the way I'm looking at this curse. Mentally you're telling yourself, not again, not again. You're in denial to be honest. You know what you're feeling. You feel the pain, you feel the aches, you feel all of it, but you're telling yourself, no, it's not happening, I'm fine. If you believe in God, you're calling on God. You're calling on someone. Mentally, you're trying to figure out what you did to deserve the pain that you're in in that moment.

Renneth's Textural Description

Renneth is a professional woman who works in the healthcare industry. She reported having a lot of trouble with her SCD, and needing her husband to sometimes help her. She described her experience with SCD patient as:

> Well, it takes on a few different forms for me. The first one is the pain, and normally I just get it in my lower back and when I turned probably around 18 or 19, it started going from just my back to my hips, and my knees, and my ankles. So there would be some mornings or sometimes where the pain is so bad that I can't really move, I can't get out of bed or if I do try to get out of bed it feels like I'm stuck or it looks like I'm stuck and I have a hard time walking around so much. Then there are other days, like I said before, where it's like I either can't move in the bed and if I have to really pee badly I have to have my husband move his leg so I can roll over to get to the bathroom. And then sometimes it gets to the point where he actually has to lift me up because I don't have the strength because of the pain to actually get off the toilet myself.

Renneth's Structural Descriptions

Renneth reported having crises about six to seven times in a year. She said she had recently been able to stay away from having frequent crises because of her medication, hydroxyurea. She explained her experience during crises as below:

> And it's hard to go from being a very active person to being stuck in a bed or being stuck at home for a while. And then I feel like when is it going to end too? I also don't know how long it's going to be there for. And I don't like to have to take pain medication, but at those times I have to and it's an internal battle with myself because I need the pain medication, but you don't want to take the pain medication, but I am smart enough to know that I have to take the pain medication. So it's not a battle that lasts too long. Thank goodness.

Shelly's Textural Descriptions

Shelly discovered she had SCD when she was a child when her parents enlightened her regarding the condition. She explained her experience having SCD as:

> It can make you sometimes feel like you're not able to do what others are able to do even so as you get older, especially if you have chronic crises, it can stifle your lifestyle and what you're trying to live like or do. Sometimes you just can't do what everybody can do because you're afraid, you don't want to catch a crisis, or you don't want anyone to look at you or certain kind of way. You try to have a sense of normalcy because when you're getting sick. People asking you all the time what's wrong? And you say, "I'm sick." And it's like that people are like, "You're always sick." Sometimes that could be very hurtful because you don't want to be known as the sick person or the sick child.

Shelly's Structural Descriptions

Shelly is a young lady, a professional who works for the port authority. She reported having crises at least five to six times per year. Shelly described her experience during crises as below:

> I've had crises in my legs, where I couldn't even move to get up to walk anywhere. It was so painful, it just was crippling. It's like I had to crawl to get on the bed or crawl to the couch. That pain could be very painful sometimes depending where the pain is, it's the different types of pain you may feel. It could be very depressing. Sometimes in stages, you get some form of depression, you feel like an outcast, so you try to develop that normalcy, but when it's going on it's

> sometimes hard, so you try to maintain your regular blend. It is not like anything other, it's not like a normal chest pain, is not like a normal back pain. It's truly sometimes it's on a thousand. Because to anybody else, they get a chest pain, it's just a little ache, but we get a chest pain and it's like you can't breathe, you feel someone stabbing you. It feels so tight and you just feel like you can't move.

Tammy's Textural Descriptions

Tammy is a young lady with SCD who explained that SCD has limited her in many areas of her life. She further explained as below:

> Well my life is affected by sickle cells because, well, you really cannot hold down a perfect job, because then you are always in the hospital. Nobody wants to put up with that type of person, worker, especially because you're unreliable. So nobody wants that relationship, no man wants a sick person either, a sick girlfriend, a sick wife.

Tammy's Structural Descriptions

Tammy spoke about having two to three big VOC that send her to the ED and eventually lead to multiple hospital admissions per year. She also expressed having a lot a little VOC that can be manager at home. She further described:

> Sickle cells pain is a pain that not even your worst enemy you want to feel the way you feel on the day you're in crisis. So you know much times you can't wear no clothes because your body hurts you so much. You don't know if it's the clothes that is bugging or if it's the blanket that is hurting, you don't know. You can't lie down when you're having pain. You barely could even stand up, much

> less to be laying on the bed thinking if you're going to die. With excruciating pain on a daily basis and you go to the hospital.

Warrior's Textural Descriptions

Warrior asserted that having SCD but is a big responsibility, depressing and stressful. She explained having SCD has restricted her in some ways. She explained that SCD but has made her to be more spiritual and believe in God. She further described:

> It restricts me from a lot of things. I had two hip replacements, so the rain makes me sicker. Financial-wise, it's affected my life tremendously because I actually work hard. What do I do to make my life better? Because, right now, I'm living off a $600 check and that's not enough for nobody. Spiritually, I go to church every Sunday. I met some people that actually blames the Lord, but I have to tell them that the Lord did not give us this. It was mankind who gave us this. He's trying to help us, or at least have things on here that could possibly help us. But He did not give it to us. It was mankind who created all of these diseases who gave it to us. I don't blame him for it, I don't blame him for anything.

Warrior's Structural Descriptions

Warrior described having VOC about nine to ten times per year. She described the pain of VOC as draining. She additionally described her experience with VOC as blow:

> Then, like the crisis itself is just a whole nine yards because all you get is throbbing, sharp, and aching pain, and you can't do nothing about it, but to just either take medicine or use your heating pad, and sometimes that don't even work.

> Physically, I feel drained. I cannot walk or it's just too much. It's like extra pain on top of pain if you try to walk with it or anything, so it's like you have to be in a cocoon. You just have to stay still, in order for my pain to go away. I shake.

Composite of Textural Descriptions

All study participants verbalized what it is like to have sickle cell disease. They spoke of the challenges of having to experience constant pain that is never going away. These individuals with SCD call themselves “warriors”. They described *Excruciating Pain* during VOC which can be as frequent as weekly. They vividly described *Depressing*, having to deal with SCD and its complications, and not being able to perform those activities in which they would have loved to participate because of their SCD status. They described *Depressing* during VOC as very serious even though many people including some doctors do not understand this. They expressed the challenges and frustrations of *Feeling Helpless* from either family members or friends during VOC to help them physically, emotionally or financially.

They also pointed out their frustration of *Dreading Healthcare Workers’ Attitude*. They expressed their fear of stigmatization from healthcare workers, especially in the ED. Some of them reported being nicknamed as 'drug seekers' by healthcare workers who either don’t have adequate knowledge of the disease or concerns regarding their illness. Through it all, participants expressed hardships and complications that are associated with having sickle cell disease. They enthusiastically spoke about what keeps them going and how the disease has made them better and stronger persons than they could have been without the disease.

Composite of Structural Descriptions

Sickle cell disease individuals in this study have all experienced VOC several times, ranging from two to more than 60 times per year. This group of people found VOC to be extremely hard to deal with. Most of them declared that VOC was tough, difficult, and that it made them miserable. Regardless of the frequency of VOC, participants spoke of *Excruciating Pain* during VOC. They eagerly described *Excruciating Pain* as intensive, unbearable, sometimes indescribable, and extremely agonizing. All participants also expressed *Depressing* during crises, as friends, families, and even some physicians do not understand the magnitude of their pain. They explained that they feel depressed and isolated during VOC because they realized that the pain would never go away and that they would have to deal with this agonizing pain for their entire life. Most participants in the study also expressed their *Feeling Helplessness*. Most participants said they always wanted someone beside them during VOC, especially their close family members. Some said it did not matter who was at their bedside, as long as there was someone beside them during this crucial time of VOC. They all believe that since the pain is usually incapacitating, they will always need help from someone to either help them move around or to get things for them.

Finally, majority of participants expressed their frustration with healthcare workers especially in the ED. They expressed *Dreading Healthcare Workers' Attitude*. Some of them pointed out that they had been given several names such "drug seeker", "drug addict", "Liar", and "frequent flyer". Some participants asserted that some physicians had refused to medicate them for pain and had sent them home in this severe pain of VOC. Some participants said they would rather go to sickle cell clinics or even

urgent care centers for VOC instead of the ED just to avoid the negative attitudes of the healthcare workers and the stigma that comes with VOC.

Synthesis

The lived experience of adults with SCD during VOC includes, *Excruciating Pain, Depressing, Feeling Helpless, and Dreading Healthcare Workers' Attitudes.* The participants explained how having SCD is physically, financially, and emotionally draining. They explained how the pain of VOC is difficult, tough, and unbearable. Regardless of the frequency of VOC, participants spoke about *Excruciating Pain*, the most common complication of vaso-occlusive crises.

These SCD persons spoke about *Depressing* during crises. They explained that during VOC, they feel down and alone, they feel like giving up on everything. Some expressed the feeling of just wanting to die in order to get rid of their suffering during VOC. They described feeling sad for a sickness they had not asked for and had not done anything to deserve.

The participants described their *Feeling Helpless* during VOC. Some participants reported not being able to move or ambulate to the bathroom during VOC therefore needing help from someone who will be by their side at all times. Some participants reported feeling helpless because of the intensity of their pain, avascular necrosis of the joints, and inability to move some part of their bodies. The participants described the challenges of facing healthcare workers especially in the ED. They fear stigma and stereotyping. They described avoiding the ED and preferring to go to sickle cell clinic or any other place that will care for them without any biases or judgmental attitudes. Some of them said they would rather stay home and manage their pain because they are afraid

of going to the ED where healthcare workers might judge them because of their pain. Through all the challenges, participants expressed being stronger because of their SCD, and that they are reminded of how delicate life is. The participants said they are grateful for being alive and being able to manage their disease as most of them have been told that their life span would be very short, and some were told that the disease could have claimed their lives.

The Theory of Self-Care of Chronic Illness

The middle range theory of self-care of chronic illness may be linked to the main themes of this study, which include *Excruciating Pain, Depressing, Feeling Helpless, and Dreading Healthcare Workers' Attitude.* The theory of self-care of chronic illness was developed by Barbara Riegel, Tiny Jaarsma, and Anna Strömberg (2012) which they derived from Dorthea Orem's grand theory of self-care. The key concepts include self-care monitoring, self-care maintenance, and self-care management. The theorists mentioned the factors that influence self-care, which includes motivation, experience, skill, culture, habits, confidence, functional and cognition and support from others, and access to care. The participants' experiences described in this study can be linked to the concepts associated with the theory of self-care of chronic illness. This connection will be further developed in Chapter Five.

Chapter Summary

This chapter presented the findings of the lived experience of adults with sickle cell disease during vaso-occlusive crises. Moustakas' (1994) transcendental phenomenology was utilized to explore the phenomena being studied. A description of each participant was outlined, and demographic data was summarized. Themes,

individual textural and structural descriptions, composite textural and structural descriptions, and a synthesis of the experience were provided. Finally, the essence of the experience was linked to the Theory of Self-care of Chronic Illness. Chapter Five will explore and interpret findings and show the connection between the themes of this study and the construct of the Theory of Self-care of Chronic Illness, the significance to nursing, strengths and limitations of this study, and recommendations for future research.

CHAPTER FIVE

DISCUSSION AND CONCLUSION OF THE INQUIRY

The purpose of this qualitative, heuristic phenomenological study was to explore the lived experience of adults with sickle cell disease during vaso-occlusive crises. This chapter presents the exploration and interpretation of the findings of the study, including themes, individual textural and structural descriptions, and a composite of overall textural and structural descriptions. The themes were related to the theory of self-care of chronic illness and a synthesis was provided of the essence of the experience as they connected with the literature. This chapter also presents the significance of the study, implications for nursing education, nursing practice, nursing research, and health and public policy. Finally, the strengths and limitations of this study are discussed with subsequent recommendations for future research.

Exploration and Meaning of the Study

Sickle cell disease is a hereditary disorder that affects people of African descent, Middle East, Indians, and Latinos. It affects approximately 100,000 of Americans (Smaldone, Findley, Manwani, Jia & Green, 2018). Sickle cell disease is associated with severe pain, chronic anemia, and organ dysfunction, leading to numerous acute and chronic complications, high healthcare utilization, a poor quality of life, and premature mortality (Smaldone, Findley, Manwani, Jia & Green, 2018). In this phenomenological study, the lived experience of adults with SCD during VOC was explored. Through the analysis of the participants' interviews, four themes were developed and presented in Chapter four. Without an understanding of the lived experience of adults with SCD

during VOC, lack of adequate knowledge of the disease, its physical and psychological complications, and the stigma associated with the disease will continue to occur.

Moustakas' transcendental phenomenology was selected as the appropriate method for data analysis because it focuses solely on the experiences of the participants and is a systematic approach ideal for novice researchers (Creswell & Poth, 2017). Participants in this study voiced their experiences through semi-structured interviews. Four themes, *Excruciating Pain, Depressing, Feeling Helpless*, and *Dreading Healthcare Workers' Attitude* emerged from the data. Individual textural and structural descriptions were established to develop the overall composite of textural and structural descriptions and the subsequent overall essence. The themes were connected to the theory of self-care of chronic illness.

Interpretive Analysis of the Findings

The experiences of these sickle cell disease participants during vaso-occlusive crises are expressed vividly in the words of this poem:

This is My Life

Born into a life of recurring bouts of pain
But this is no ordinary pain-
It is the same kind of pain
From the same exact cause
Experienced by someone undergoing a heart attack
With no scientific way to measure the agony caused by the pain.

This is my life

Frail, fragile and under grown

Cannot keep up with my peers on the playground

Have to dress heavily all-day round

So I can stay stress-free all year round

This is my life

“Him again?!”

“What a Wimp”

“Here we go again”

Is what they say

At the sight of me, at the gate.

“I think he’s faking”

Is what they are thinking

When I am seriously hurting.

The wall of doubt I run into

When help is all I am running to.

This is my life

My chances are slim

Full adult life I may not get to feel

Band aid is all what they get to give

This is my life

My chances can be real

Possibilities big

If the doubts will cease

And the care and attention increase with a balm of love

The pain will ease if rubbed over to soothe

This is my life – The Sickle Cell Child (Sickle Cell Warriors, 2015).

The participants in this study had various experiences having VOC of sickle cell disease. Despite differences in age, gender, educational level, occupation, and number of VOC per year, all participants spoke about their various challenges and difficulties during VOC. Through the process of data analysis and reflective journaling, the main simultaneous themes of *Excruciating Pain, Depressing, Feeling Helplessness,* and *Dreading Healthcare Workers' Attitude* emerged and yielded the individual's textual and structural descriptions, the overall composite of textural and structural descriptions, and the final synthesis of the essence of the experience.

Theme: Excruciating Pain

There are limited studies and literature on the pain of VOC. Carvalho, Santos, Izidoro, Caldeira dos Santos, and Batista Santos (2016) explained that pain is the hallmark of SCD and that it dominates the clinical image of individuals with sickle cell disease throughout their lives. They further expound that the nature of the disease is unpredictable and can be precipitated by known and unknown factors, and that it is the most common cause of more than 90% of hospitalizations leading to recurring visits and

hospitalizations in the ED. Coleman, Ellis-Caird, McGowan, and Benjamin (2016) wrote about the "unimaginable" and "indescribable" pain of VOC (p. 190). The participants in their study described their pain as "unimaginable" and "indescribable". Some described their pain as worse than labor pain. Some personified the pain because they confirmed to have been living with it for their entire life.

According to Fosdal (2015), the main symptom of SCD is pain related to vaso-occlusion. In this study, it was reported that the pain of VOC is distinct from other pain and can be excruciating, and has more intense than pain associated with surgery. In the study, it was reported that VOC pain felt like two sides squeezed together with knives and hammers penetrating from both sides. In addition, it was conveyed that the pain of VOC was associated with decreased participation in sports and favorite activities, increased school absence, and increased incompletion of schoolwork. The researcher listed word descriptors for pain frequently used by the SCD participants in her study: “throbbing, aching, hurting, pounding, awful, crying, uncomfortable, annoying, uncontrollable, and hit with metal bat or brick”.

Elahi, Tahery, Ahmadi, Rostami, and Elahi (2017) conducted a qualitative study in Iran to describe the lived experienced of patients with SCD during the evolution of a painful event and the solutions for controlling and managing it. The study explored the experiences of SCD patients in the management and control of pain and disease crisis in Iran through in-depth, unstructured interviews were done with the participants. The researchers explained that conservative measures such as the avoidance of excessive physical and mental stress, adequate hydration, and prescribed opioid analgesics could be used to manage the excruciating pain of SCD. The researchers asserted that the SCD

patients adhere seriously, and use to all the measures of self-care, other non-pharmacologic measures, and pharmacologic measures. However, these patients learn at an early age that they have very little if any control over the unpredictable and excruciating VOC pain episodes and complications that linger throughout their lives. Opioid analgesics are often prescribed in the pharmacologic management of VOC pain (Myers & Eckes, 2012). However, participants' perceptions of negative physician and hospital experiences and their inability to pay for the medication prompted many to manage their SCD with complementary and alternative medicine (CAM). Only 23% of the participants in this study reported the CAM options to be beneficial. The investigators concluded that since nursing plays a significant part in the pain management experience of persons with SCD, and they are usually the first providers that SCD patients come in contact with, the relationship nurses develop with SCD patients is important in helping to determine satisfaction with the patients' pain experience.

The findings from this study are consistent with the literature on *Excruciating Pain.* In the following excerpts, participants convey what their VOC pain feels like. In addition, they spoke about the unbearable pain they experience through a VOC.

In her story below, **Denise** verbalized that *Excruciating Pain* feels very uncomfortable and depressing. She said:

> It's painful. It's like the worst pain. The worst, and you get miserable, you get frustrated, you don't want be bothered. It's painful and you don't wish that on your worst enemy, I always think, because it's painful. Sometimes you can't walk. Sometimes it even hurts to move anything. It's painful, it's frustrating, depressing, miserable, and even more miserable.

KO pointed out that *Excruciating Pain* is stressful and tedious. He further described as below:

> The pain is, it feels like I'm getting stabbed everywhere at the same time. All my bones are just breaking, and it, yeah, I can't even explain it. It's crazy man, sometimes it's like my chest getting tightening up on me. Um, my stomach hurts, I feel it in my back, you know. It's very stressful man. It's hurting.

Lawa's explanation of *Excruciating Pain* is as described below:

> The pain is something like, I feel like I'm in another state of mind. Like I'm not even here because I'm just down. I'm always in pain. The pain is so bad you can't even describe it sometimes. But some people be like, "Oh, what is your pain one through 10 or what does the pain feel like?" And it's just a pain that you can't describe. It hurts so much that you don't even know how to describe it. So I'm not always in the right mind.

Cerena described *Excruciating Pain* as very isolating and depressing. She further described:

> It's almost ridiculous to try to limit it to that to explain to somebody what it feels like. With the bone crises, it's like somebody beating you with a metal bat. It hurts from the inside. The bone pain is really, really terrible. It's one of the most severe things that I've ever felt. It limits everything you can do. If you can't breathe, you're essentially dead, right?

According to **KingJeff**, Vaso-occlusive crises pain is agonizing and unbearable. He described it further:

> So, my pain when it do come, it's been very excruciating pain. It starts in my lower back and it go down to my legs. My sickle cell crisis attacks me, it attacks me from my lower back all the way down to my legs. It hits my hips also. So that it's hard for me to walk and it's hard for me to really do anything. A pain will come excruciating. It be like sharp, throbbing pain. The pain crisis I go through is very, it's very excruciating.

In agreement with the literature, the findings from this study revealed that all sickle cell patients describe the pain of VOC as severe and extremely hurtful. They all describe what *Excruciating Pain* of vaso-occlusive crises feels like.

Theme: Depressing

People diagnosed with chronic illnesses often report experiencing depression and isolation related mainly to the struggles associated with their illness. As a result, the prevalence of a major depressive episode found in individuals suffering from a chronic medical illness varies from as low as 5% to as high as 40% or higher in others (Morgan et al., 2014). Understanding and coping with the medical and psychosocial challenges associated with SCD is of highest importance throughout a person's lifespan, as the people affected were born with it as a genetic disorder. The incidence of depressive symptoms in SCD ranges from as low as 21.6% to a high of 56.6% (Morgan et al., 2014). Sickle cell disease persons suffer chronic pain of VOC, which predisposes them to anxiety, depression, fatigue, impaired physical function mobility, and worse quality of life (Badawy, Barrera, Cai, & Thompson, 2018).

Davis and Brown (2016) found that negative thinking, depression, frequent hospitalizations, and passive coping were associated with pain crises of VOC. They discussed that two of the factors that determine quality of life for SCD sufferers are anxiety and depression. They asserted that anxiety and depression are associated with low self-esteem, feelings of hopelessness due to frequent pain, hospitalizations, and loss of employment. They further explained that the severity of pain of VOC varies in each individual and can cause physical, emotional, and psychological disturbances. The researchers concluded that the combination of SCD and depression increases psychological morbidity and mortality, and that mental health and social health encompasses the quality of life among patients with SCD.

Miller-Matero, Chipungu, Martinez, Eshelman, and Eisenstein (2017) declared that there is increase prevalence of depression among blacks with chronic illness such as SCD. They explored symptoms of depression in blacks with SCD in their study, they found that patients with SCD have been estimated to have a lifetime depression prevalence at 50 percent. They concluded that there is evidence that sickle cell pain may be related to depression and suicide in patients with SCD. Carvalho, Santos, Izidoro, Caldeira dos Santos, and Batista Santos (2016), pointed out that the participants in their study described pain of VOC as persistent, always present, consistent, recurrent, and the pain that does not go away. They believe that pain of this magnitude can cause psychological changes, sleep disorders, and psychopathology as depression, personality disorders, and anxiety.

Raji, Lawani and James (2016) did a cross sectional study in southern Nigeria to determine the current and lifetime prevalence of depression among adults with SCD and

identify relevant socio-demographic and clinical correlates. The researchers asserted that the prevalence of current depression was 16.6%, while lifetime prevalence was 29.8%. they further explained that current depression was significantly associated with frequent analgesic use, unemployment, low income, low educational status, and subjective pain. The researchers declared. The study concluded that depression is common among adults with sickle cell disease and is significantly associated with severity of subjective pain.

The findings from this study are congruent with the literature. Among the many, Isolation is part of emotional and psychological reactions associated with the pain of VOC. The stories from the participants below display their *Depressing* experience:

Shelly described her emotional state during VOC. She expressed being depressed and frustrated from inability to do regular activities. She further described her emotional experience as:

> Having sickle cell disease, when you're going through a crisis can be very painful, depressing, and depending on where you're having the effect on your body could be the severity of the pain as far as from your chest, to your back, to your legs, to your arms, the paining. During that time, it could be very depressing. Sometimes in stages you get some form of depression, you feel like an outcast, when it's going on it's sometimes hard, so you try to maintain your regular blend. It can change that, when you're going through a lot when you're just not up to it and you wake up and you feeling some sort of pain, It's like, it's going be a tough day and you're not able to enjoy the day as you would hope to.

Cerena spoke about *Depressing* during VOC. She stated:

> It's very isolating and depressing. It's lonely. We don't really talk about the psychological impact of it. Definitely depression related to sickle cell, related to chronic illness. Of course, the pain. I perceive more kindness from strangers in certain cases. It's just that kind of mitigates the loneliness and isolation that you feel when the crises start to come on. You become depressed.

Shelly expressed her emotional state during VOC. She described:

> During that time, it could be very depressing. Sometimes you just can't do what everybody can do because you're afraid, you don't want to catch a crisis. Sometimes in stages you get some form of depression, you feel like an outcast, so you try to develop that normalcy, but when it's going on it's sometimes hard.

K.O explained *Depressing* during crises is very serious especially that many healthcare workers had told him he might not live long. He further described:

> It is depressing. But then again it does make you want to, you know, it does make you feel like a better person. You know because I want to become a better person because this disease, there's only certain things that we're limited to do. And every day is like a test, some people say we're not supposed to live past 25 or something. I'm 26, and I'm still here so it's like, um, an accomplishment for me. I feel like every day is an accomplishment. Like I'm overcoming all the negativity and all the other stuff. It's yeah.

Ladybug explained her feelings during crises especially when she has to be admitted to the hospital. She states:

> There are times where, you really sit and think that it's hard to be in pain 24/7, and you're not with your kids as you should be. In the hospital and away from them. It's kind of depressing a little bit. You can't really let it get the best of you. You got to keep going, that's why I keep a picture of my kids as my screensaver because they keep me going.

Lawa described her state of mind during VOC. She states: “I will always say that emotionally, I'm always down. I'm never happy. I'm always crying. I always have an attitude. I don't want nobody talking to me. I wouldn't talk to people. I just isolate myself from everybody.” **Brad** verbalized his experience during VOC. He said “So, I try to stay healthy and do my research and sometimes it's tough to research it because you start finding things about other people, like life expectancies, it just depresses you.”

Warrior conveyed what *Depressing* felt like during VOC. She described her experience as below:

> It's like a depression sometimes or a stress because you're actually stressing over your sickness, which causes you to get sick. Then, like the crisis itself is just a whole nine yards because all you get is throbbing, sharp, and aching pain, and you can't do nothing about it, but to just either take medicine or use your heating pad, and sometimes that don't even work. I'm okay mental-wise, but sometimes I do have a depression type of thing. But one thing I do is pray to God and then I get myself back into the energy that I need to be in. Mental-wise, I feel like sometimes I cannot do what I need to do for myself and I don't like other people doing it for me.

The findings from this study are consistent with the literature. Many SCD individuals experience feelings of depression and isolation during VOC though individual reasons for feeling depressed and isolated might be different.

Theme: Feeling Helpless

Matthie and Jenerette (2015) declared that the pain of VOC can last for months or years and is accompanied by helplessness, suffering, anxiety, despair, depression, insomnia, and loneliness. Keane and Defoe (2016) explained that the needs of SCD patients are complex. In their study, families of SCD patients reported that their own needs were sacrificed, and that this had a knock-on effect, which exacerbated all other stresses within the family. Brown et al. (2010) explained that caring for an individual with sickle cell disease poses extra demands on caregivers, both practically and psychologically, which may impact on the health and well-being of their caregivers and family members. Despite help from families or other caregivers, the participants still described *Feeling of Helpless.*

Amara, Almeida, Santos, Oliveira, and Lanza (2015) conducted a descriptive quantitative study to describe socio-demographic and economic characteristics, lifestyle, clinical manifestations, use of medications and monitoring of adults with SCD. All of them reported painful crises and fatigue. All the participants reported having painful, incapacitating pain crises that impaired their ability to carry out daily activities. The researchers discussed that the participants in their study reported that the frequency of their painful crises contributed to their feeling of helplessness. Only very few of their study participants reported that they had the support to carry out their daily activities during painful crises. About 95% reported they did not have any support during these

incapacitated crises that left them helpless. All participants reported fatigue throughout the year, and the intensity of the fatigues was moderate. The researchers concluded that the implications and the complications of SCD could be mitigated through primary, secondary and tertiary health care, according to the needs of those adults with the disease.

KO expressed that he would rather have a family member or a friend beside him during crises, as that uplifts his spirit. He said:

> I mean yeah, during crisis, family, it helps because then it shows that you have support and people that care about you. It really does take a lot off of you, cause when you in crisis, and you have nobody around you, it kind of makes you sad. You know what I mean, like, stressed out more. And then you feel like, well you have family numbers here with you, I mean you still in crisis and it hurts but, your spirit is uplifted.

Madden Girl declared she needs people around her during crises, especially her mother. She further explained:

> I need family. I would say that. Family, and friends, just having somebody be there for you is very important regardless of what you're going through. No matter what it is, you need people. That's what we're here for, it’s to be there for each other in our times of need. I need my mom, because I've always had my mom from the very beginning. It's been me and her. So that's what I'm accustomed to. As I got older, I thought I could do it on my own. I actually tried to do it on my own. No, it wasn't working out at all.

Jaysmoove expressed his needs for his family and their support at the time of VOC. He further described his need as:

> Every time the episodes do happen, I do actually need to be ... I actually need just people encouraging me. I need family with my mom. It just makes me happier. It makes me want to actually just ... I don't know. I feel better and more at peace whenever I have people come to me, around me, comforting me just making sure I'm okay. Just positive energy around me whenever that does happen. So whenever I do have a crisis I just need just everything to be flowing smoothly.

Shelly explained that she feels helpless during VOC but does not want anyone to worry about her during the crises.

> You feel helpless and sometimes you feel really defeated and once you get into that point… The few times I've been in the hospital, I more or less felt more of a helpless, depressive spirit because of that simple fact that is like, I don't want anyone worrying about me, I don't want anyone worrying, "Is she okay? What's going on?" I don't want the doctors or nurses looking at me like, "*She's just helpless, just give her this*".

Tammy verbalized that she needs support during VOC even though it could be difficult for people to deal with her during crises. She further explained:

> Need the support from my parents. So that's the honest way I get through some of my crises, it's through my mom and my family. I don't have my dad's support all the way but then my mom is the biggest. So, yeah it be hard sometimes, but I try

> to make it, make through the day, make through my pain. So, go through it together with family, if you have.

Denise reported that having support during VOC is a good thing as it makes her feel less helpless and more comfortable. She further described:

> Yeah. And I get that from my family. My mom, my brother, my sisters. They'll call me if they can't come or they'll come visit me. But sometimes, I'm not gonna lie, they don't want to deal with me sometimes because when I'm in pain it's aggravating, and they just don't want to deal with that, so, they'll wait a couple of days before they will come see me. We'll still talk, it's good to have that support. It's good to have that support because it's hard.

Theme: Dreading Healthcare Workers' Attitude.

Reich, Cantrell, and Smeltzer (2019) conducted a literature search over several years on SCD individuals transitioning to young adults. The researchers declared that persons with SCD may be stigmatized by healthcare providers and communities, and that this can have a negative effect on their physical and psychological health. They further declared that healthcare providers' negative attitudes can affect care and that providers in general have negative attitudes toward patients with SCD. The study concluded that stigmatization can make patients and families to distrust healthcare providers. The researchers suggested ensuring that nurses and other healthcare providers are educated about SCD as that is paramount to treatment success and decreasing patient morbidity and mortality. Providers' attitudes add to continued healthcare disparities for SCD sufferers. Healthcare providers' attitudes should change in order to help improve racial

health care disparities facing SCD sufferers. Though not the sole issue, healthcare providers' attitudes must change in order to help improve health care disparities (Nelson and Hackman, 2013).

Bergman and Diamond (2013) in an article entitled *Sickle Cell Disease and the "Difficult Patient" Conundrum* examined the relationship between the "difficult patient" conundrum and sickle cell disease. The authors explained that most SCD patients view sickle cell pain as more intense than postoperative pain but as severe as the pain of cancer. Some patients believe that the pain of sickle crises dominates their lives. Some consider sickle cell as pain and suffering. They enlightened that VOC is the main reason SCD patients visit the ER. Since pain is mostly subjective, there are only a few measures-such as the patient's vital signs, to determine whether, and to what extent, a patient is in pain. Besides, although SCD sufferers complain of serious pain, they frequently involve in activities such as watching television and talking on the telephone which are inconsistent the old picture of patients in severe pain. When physicians note or perceive these discrepancies between patient behavior and their self-diagnosed pain score, it further mixes issues of trust. Principal among causes of mistrust between SCD patients and healthcare providers is the misleading belief that sickle cell patients are "drug seekers" or are addicted to pain medications.

The authors argued that despite an evident medical need for the administration of pain medications, some healthcare workers' belief that SCD patients are drug seekers has not changed. They claimed that most ED physicians and hematologists believe that more than 20% of sickle cell patients were dependent on analgesics, and so are uncomfortable prescribing opioids for them. They further enlightened that SCD patients report negative

health outlook and medical care opinions based on previous experiences with the healthcare system. Sickle cell patients reported being treated rudely and experienced delays in care during VOC. Some participants reported that they avoided the ED until they extremely needed utilization. Suspicion and perceived drug dependence combine to negatively influence sickle cell patients care in the clinical setting.

The authors concluded that the undertreatment of pain during VOC is often associated with health care disparity issues and that pain complaints of racial and ethnic minorities are less likely to receive adequate attention. The beliefs and attitude of ED medical professionals are critical in determining the quality of care that a SCD patient receives.

John Martin expressed his frustration of being stereotyped and *Dreading Healthcare Workers' Attitude* during crises especially during ED visitations. He explained:

> It's just a hassle, everyday struggles that people don't know about. People can't see. But people always want to judge you off of what, what you are telling them and all that. It's just a hassle. You at the ER, you just need a doctor to open his ears, to be open minded, to listen to what you, what you trying to tell them that's going on with you. Because sometimes they don't even listen. They just, okay, he's in pain, and he's a drug addict or let's just try to get him out of here and stuff.

Warrior described that she gets better care in sickle cell clinics than the emergency departments. She described further:

> But, in the ER and in the hospital is horrible because I'm not, I'm not sure if the new material that they use to teach the nurses or doctors that's coming on, or the

doctors that's already seeing us as drug addicts is teaching the ones that are coming on. Everyone, not everyone, it's probably like a good 10% that's not, but mostly when you go in there, it's just that they think you're a drug addict and they think you're coming in for meds. It takes you more and that stresses you out more, which causes more pain because stress with sickle cell patients makes your pain worse. They make it more stressful in the ER or in the ER room. If I can't move and the clinic is closed, I will definitely go to the ER.

Tammy explained her frustration and *Dreading Healthcare workers' Attitude* during ED visits. She described her experience as below:

Well the care that you get from nurses, doctors, especially the doctors is, "you're here for drugs". The first thing they see sickle cell disease on your paper is, you're here for drugs. So they come to you as you are a druggie. So they don't come to you nicely. They'll come, "Oh yes, what's wrong with you today?." You explain, by the time you tell them the real problem you here for, they already cut you off and leave the room. And then they give you a couple of hours in the ER and then they discharge you. So they give you one hour, two hours in the ER, they give you something for pain. You're still in pain but they discharge you anyway. So the care that you get is not a welcoming care from nurses and doctors. Nurses take their precious time to come give you the medication. You could be balling down the place. You could die and come back alive and the nurses still didn't reach your room. So, they don't treat you as a human being, they treat you as a druggie in the ER. A lot of sicklers die because of the result of how they think you are. So they don't give you the time and the treatment. They don't spend the time with you. So,

> a lot of sicklers die as a result of it. They keep you in the ER three, four hours, in the emergency room before you can be seen. So, unless the labs result is matching up to what you saying, they don't give you no help.

Madden Girl shared her experience of being judged negatively by healthcare workers during crises. She explained:

> You go to the hospital; you say you're having a sickle cell crisis. You have to cross your fingers and honestly pray that you're lucky enough to get a good doctor, a compassionate doctor, one that's not going to judge you and is not going to overlook you because of the color of your skin and because of what this, the stigma that's behind sickle cell disease, period. You'll have doctors that will say to you, you're fine. Haven't done any blood work. They just look at you and they say, well, you're not the floor crying. You're not screaming. How can you possibly be in pain?

Shelly described her frustration during ED visits for pain of VOC. She explained that healthcare workers fail to understand the pain of VOC, and as a result, do not treat SCD patients according to the level of their pain. She further said:

> I would say the care is minimal and for sickle cell patients, it can't be minimal. It needs to be intense and it needs to be approached in a different manner. Because as I mentioned, it's a different level of pain. You're asking me one to rate my pain to 10 but my pain is a 20 and it's because it's not a normal chest pain, it's not a normal back pain, it's not a normal arm pain, it’s an excruciating pain, beyond anyone's truly understanding. It's an intense pain that is going through your body.

> Sometimes the medicine they give you, if you're a chronic person, doesn't work. I think they fail to realize that. Sometimes when you're like, I need more medicine or I need a different one, they're thinking you're just trying to search for a drug or get more drugs. Having them understand the pain is very difficult because they don't want to listen to the patient because they have their medical degrees and backgrounds. So they think they understand it all and sometimes they really miss the ball with that, treating the level of pain that a sickle cell sufferer experiences.

Every participant conveyed *Dreading Healthcare Workers' Attitude* during Vaso-occlusive crises. Some explained their frustrations during ED visits. Some expressed being reluctant visiting the ED because of their fear for healthcare workers' negative attitudes towards them during their pain crises. The findings from this study are consistent with the current literature.

Connection of the Theory of Self-care of Chronic Illness to Themes

In combination with this current study's findings, Riegel, Jaarsma, & Strömberg, (2012)'s middle-range theory of Self-care of Chronic Illness may be linked to the overarching themes of *Excruciating Pain, Depressing, Feeling Helpless,* and *Dreading Healthcare Workers' Attitude.* Twenty SCD individuals who participated in this study described what it means having SCD and dealing with VOC and its complications. Their eagerness to share their experiences allowed for a deeper understanding of what it means to be in VOC. This researcher linked the findings of this current study with the theory of Self-Care of Chronic Illness. Riegel, Jaarsma and Strömberg (2012), developed this theory addressing the process of maintaining health with health-promoting practices within the context of the management required of a chronic illness. The key concepts of

the theory include self-care maintenance, self-care monitoring, and self-care management. The concepts of the theory might help individuals with SCD control and manage excruciating pain, depressing, feeling of helpless, and dreading healthcare workers' attitude. The theory was derived from Dorthea Orem's grand Theory of Self-care.

The theorists define self-care maintenance as those behaviors used by patients with a chronic illness to maintain physical and emotional stability. They explained that self-care monitoring refers to the process of observing oneself for changes in signs and symptoms, and that self-care management is the response to signs and symptoms when they occur. Some of the assumptions of this middle range theory are:

> (1) Mastery of self-care maintenance precedes mastery of self-care management because self-care maintenance is less complex than the decision-making required of self-care management.
>
> (2) Self-care monitoring for changes in signs or symptoms is necessary for effective self-care management because one cannot make a decision about a change unless it has been noticed and evaluated.
>
> (3) Individuals who perform evidence-based self-care have better outcomes than those who perform self-care that is not evidence-based (p.13).

The theory also establishes that there are factors affecting self-care which are experience and skill, motivation, cultural beliefs and values, confidence, habits, functional and cognitive abilities, support from others, and access to care. These attributes have been seen to affect self-care in SCD individuals.

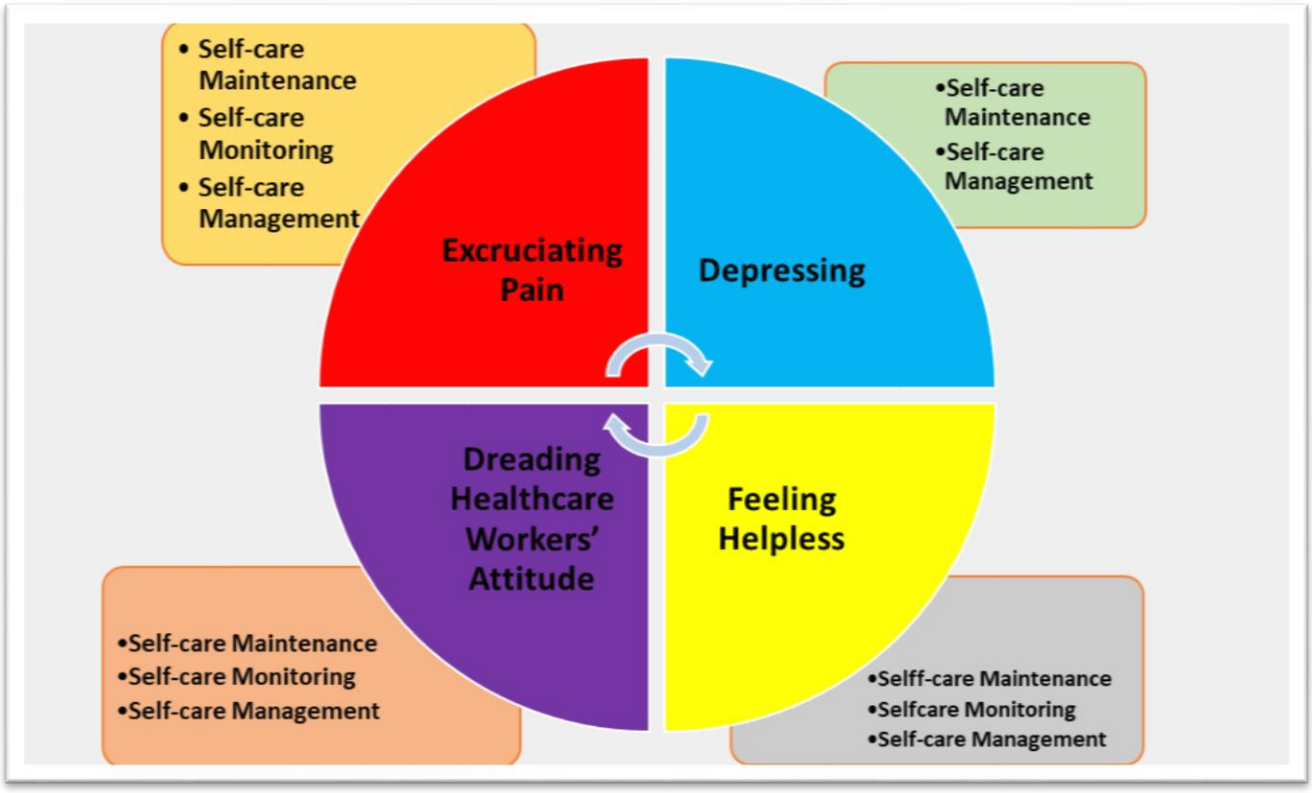


Figure 4: Theory of Self-care of Chronic Illness and the Voices of Individual with VOC. (Adeagbo, 2019).

Self-care Maintenance

Self-care maintenance is consistent with current literature. Riegel, Jaarsma, and Strömberg (2012) define self-care maintenance as those behaviors used by patients with a chronic illness to maintain physical and emotional stability. They further explained that these behaviors may be entirely self-determined or reflect recommendations that are jointly agreed on between patients and their healthcare provider. Audulv (2013) described self-maintenance as the approaches a person undertakes to control disease, promote health, and live well with illness. Audulv (2013) asserts that individuals' self-maintenance is a vital part of the care of people with chronic illness, and that most care of

chronic illness is performed by people in their own homes. All people are influenced by their social networks and health-care providers. Self-care maintenance also connects with the themes of *Excruciation Pain, Feeling Helpless, Depressing,* and *Dreading Healthcare Workers' Attitude.* All participants in this current study verbalized the importance of self-care maintenance in order to control the frequency of their vaso-occlusive crises. The participants explained that self-maintenance is significant for them as SCD sufferers because it helps them minimize the frequency of VOC and its associated complications of pain, depression, and isolation, helplessness, and stigmatization from healthcare workers.

Lawa spoke about the importance of self-care maintenance in minimizing the frequency of her VOC. She states:

> I know how to handle my health better, like the importance of drinking water. I know if it's raining, I have to cover myself up or I can't be in water, or certain things I shouldn't be eating. I stopped eating junk food at the age of probably 13, because I knew it had an effect on my sickle cell. I won't eat out. So that's how you know I know how to cook. I won't eat out. I won't eat McDonald's, I won't eat Burger King, I won't eat any of that because I knew it had an effect on my health.

Brad explained how he keeps himself healthy and maintains himself to avoid vaso-occlusive crises when possible. He described:

> I mean now that I'm older, I first make sure that I do cardio when I go to the gym to keep my stamina up and it's not even as bad as it used to be back then because I do a lot of things. I know in the back of my mind that that's there and if I don't

> stay healthy and workout, it can go bad again. I also try to make sure that I'm always healthy as possible, like within my physical fitness and eating healthy. It's kind of hard to eat healthy when you're on a college budget and you're young and everything looks so tempting and you can just eat it all. I try to be healthy because since I know I have this disease, I don't want any other things to creep up on me, so I have to deal with sickle cell and this and this, like this is already too much to handle. So, I try to stay healthy and do my research.

Jaz verbalized maintaining herself holistically to help avoid having multiple VOC and visiting the ED. She said:

> With my life, I can't be in a wheelchair. I can't be confined and cooped up, so that's why I keep going whether or not it's I'm hurting, I'm in pain. I will find myself outside walking, doing something just to be active. Honestly, I would do it all holistic, the yams and all the different kinds of foods and stuff.

LadyBug spoke about self-maintenance and the importance of being consistent taking her medicine.

> I'm trying to learn how to control my stress level. That's my biggest downfall that I'm still trying to get control after all these years. It's really hard, but I'm getting the hang of it slowly. Really all I do is be consistent with the medicine. And you stay on a schedule and you stay consistent and always on time, or early. It controls my pain a lot quicker. So I get in and out of the hospital a lot quicker. And mentally I'm good, you just keep me happy, be nice to me and offer anything that'll help ease my stays and get back to my kids. Also trying to hydrate myself to control my crises. Hydrate, I'm working on. I really need to work on that more.

> But, as in stress, it's not that easy to stay away from. But, I'm working on controlling

Cerena explained how she self-maintenance has helped her stay away from the ER. She said:

> Okay, so because I've been able to use other therapies, I've been able to cut down my crises and cut down the amount of pain that I experience. Before that, I would be in the hospital couple times every month. Now, I think I haven't been in the hospital almost a year because of the therapies and things that I use outside of- Right, so the research that I've done on sickle cell shows that it's the pain and the issues that come up with it are nutrient related, so your body isn't getting enough nutrients and hydration and all that, so I take sea moss, which is algae and algae produces 70% of the oxygen on Earth, 70 to 80%. Yeah. I use chlorophyll which is the green pigment in plants as a supplement in water to help with oxygenation as well. I use molasses. Molasses has a lot of iron and so molasses is what they use to produce sugar, but by the time it gets to be white sugar, it's so depleted. Right, right. When you use molasses and the dark black strap molasses, it has a lot of nutrient dense. It's very nutrient dense because of how deep the sugar cane goes into the ground. There's high iron content.

Self-care Monitoring

Self-care monitoring is defined by Riegel, Jaarsma and Strömberg (2012) as a process of routine, vigilant body monitoring, surveillance, or “body listening” (p.6). Patients who are well-informed are more capable of monitoring their health, coping with their illness, and adhering to treatment. The goal of self-care monitoring is recognition

that a change has occurred. When signs and symptoms are detected early and the seriousness of them is understood, action can be taken before the situation escalates Riegel, Jaarsma & Strömberg (2012). Self-monitoring is a component needed to optimize the effectiveness of self-management interventions. These interventions are most effective in reducing healthcare usage. Self-monitoring in the context of chronic illness has been defined as the patient undertaking one or more activities such a self-measurement of symptoms, vital signs, behavior or psychological well-being, treatment, lifestyle or help-seeking behavior as a result of self-awareness (McBain, Newman, & Shipley, 2015).

Self-monitoring connects with the themes of *Excruciating Pain, Depressing*, and *Dreading Healthcare Workers' Attitude.* Participants in this current study spoke about monitoring themselves, modifying their health behaviors and lifestyles so their pain of VOC, feeling of depression, and having to visit the ED will be controlled and reduced to the minimum.

Shelly verbalized how she monitors herself so she could avoid VOC and its complications of pain and depression, and minimize ED visits. She said:

> But you have to be cautious and honestly that's something that I try to do because you have little pain and I just started rubbing myself to make sure to try to avoid a crisis. So it's being very self-aware and cautious on a day to day basis. when I see myself falling into that sad, depressive state, I just remember that God is with me always praying and asking for healing.

Warrior explained that she monitors those factors that can precipitate VOC and

tries to avoid them as much as possible. She states:

> It restricts me from a lot of things because I took it upon myself to see what's my restrictions with sickle cell because some people could get in the pool, some people can't, or the weather changes. With me, because I had two hip replacements, I try to avoid rain, the rain makes me sicker. My hips would know if it's starting to rain, so it'll throb a little bit and now since I just got caught in the rain also, it just make it a little hard. I feel a little pain coming on, but I'm fine. Other than that, I don't know. I learned to live within it, but also don't exceed too much necessarily. As in, I would do things that I know I can do, but I'm not going to exceed it to what I'm not going to go into a crisis. Because in the pool, I have to go on there for 10 minutes, get out, dry up, and then go back in. I can't stay in there for a long time.

Brad talked about the importance of self-monitoring especially while on hydroxyurea. He said:

> You have to go to checkups, they have to monitor you. I only really had to go every month is because of the drug that they placed me on as a result of one of my crises, hydroxyurea, because it was new at the time. It's not so new anymore and it's like a chemo drug. So, every month they had to be checking up, making sure everything was still going right and everything was always going right. I don't remember a time where something went wrong because of the drug. So, that's what it's like. I think they had me on Percocet the last time and that's a highly addictive drug and because I know that, like once I feel the pain gradually getting better I'll stop taking the medicine even when they want me to take the medicine.

> I personally just don't want to be addicted to anything in my life, like I don't like to have anything control my actions and I know that people can get addicted to it. It feels nice to not be in pain, but if I feel the pain going down it gives me more ease than me trying to suppress the pain. So, I don't particularly like to get medicine when I'm in pain unless it's really bad and I can't operate.

John Martin spoke about monitoring his crises by watching the weather to avoid crises. She states:

> And then crises, they get worse by the conditions of the weather. So, you gotta monitor. Because if it's raining, you ain't doing that because you don't want to go out in the rain and get wet and then you'd be, your body's physically hurting from all this. It's just the moisture in the air. It really affects your bones and all that. So even when they get cold you know, we know when it's getting cold, we have a little pain on the ache or something. It's something that, we know the weather is changing.

Jasmoove discussed the importance of self-monitoring to avoid priapism, a complication of VOC, and stay as healthy as possible. He said:

> The thing that does help me, actually, is just staying hydrated with all the crisis. If I stay hydrated and I just keep a good diet, then I feel perfectly normal, and I just ... It just helps me to just live throughout my life and just live a normal lifetime. Ever since I cut out dairy from my diet, I haven't experienced priapism. Like completely milk ... Not eggs. Milk, butter. What else? Really anything like cheese. Yeah. I cut all that out of my diet. That definitely helped with my priapism. Priapism, actually, is a sickle cell crisis. Different things that I think just

> hydrate you, and just like get everything flowing back into your body. So that's why they tell you to stay hydrated. So if you stay hydrated, there's more room for the blood to flow. That's why I drink as much water as I can every day. Anything hydrating is a good for me, something I need. I personally feel like ... I drink more water than the average person regardless. That being said ... The good things that I do need that make it positive are water. That's why I even cut, like I said earlier, I cut everything as far as dairy, and products that just wouldn't coexist with my sickle cell.

King Jeff explained he preferred to monitor himself first during crises but would go to the ER only if the crises got worse. He declared:

> If I have pain medication at home, I'll take my pain medication at least a couple of times throughout the day. If that don't work then I'll seek medical attention, which is the ER. For me, for the past three, four months I haven't been prescribed any pain medication. if I have pain medication at home, it could vary from at least three to four or five pain crisis a year. You have to take pain medication cause when you take pain medications just to take pain medication, when you're really in crisis, it really don't do nothing for you. Because your tolerance is already is high. So, I like to take my pain medication when it's... When I was getting... When I feel like the pain is almost unbearable.

Self-care Management

Per Riegel, Jaarsma & Strömberg (2012), self-care management involves an evaluation of changes in physical and emotional signs and symptoms to determine if action is needed. These changes may be due to illness, treatment, or the environment.

They further explained that self-care management requires attention to the efficacy of a treatment to evaluate whether or not that approach should be tried again in the future. Cutler, Crawford, and Engleking (2018) defines self-management as strategies that allow patients to gain confidence and meet their personal care goals is important for improved health and reduction in healthcare use. Knowledgeable, confident patients who practice self-management experience improved health and use fewer healthcare resources. Self-care management connects with *Excruciation Pain, Depressing, Feeling helpless,* and *Dreading Healthcare Workers' Attitudes.* Participants in this current study spoke about the importance of managing themselves during VOC despites its complications of pain, helplessness, depression, isolation, stigmatization and stereotyping from healthcare workers.

Keisha spoke about managing herself during crises. She reported reevaluating her need for an IV access (port) during one of her crises. She further explained:

> Yeah, and then with the port, they keep saying, "You need another port." I'm like, "No, I don't. I'll be just fine." This is the funny part, one time, I had to actually call out a nurse on something she was saying because I don't have a port. So, she's like, "Oh, you know if you had a port, the med would work faster." I said, "No, it wouldn't. I would just get a rush faster." So, she looked at me. I was like, "Yeah", like don't do that. And ports cause a lot honestly. I see girls I've met, and they're going through the sickle cell clinic. Their ports get infected today while they're in surgery having a new one installed, and I'm like, "Why? Your body still has that infections in there. Why not give it six months and then get a new one put in?" And that's what I've done with the three ports that I had. They got infected. Six

months later, I would get another one, but, after my third port, after the clot, I said no.

Jaysmoove verbalized that he manages himself with diet to avoid priapism associated with VOC. He further explained:

> I cut out dairy from my diet. Ever since I cut out dairy from my diet, I haven't experienced priapism. Like completely milk ... Not eggs, milk, butter. What else? Really anything like cheese. Yeah. I cut all that out of my diet. That definitely helped with my priapism. Priapism, actually, is a sickle cell crisis. The thing that does help me, actually, is just staying hydrated with all the crisis. If I stay hydrated and I just keep a good diet, then I feel perfectly normal, and I just. It just helps me to just live throughout my life and just live a normal lifetime. That's why I drink as much water as I can every day. I personally feel like I drink more water than the average person regardless. That being said, the good things that I do need that make it positive are water, chill music, just clear my mind. Water is as far as drinking water or being soaked. I would say warm water, because that makes me more uncomfortable when I'm in a hot bath I'm just soaking. That makes me feel more at peace. Like I said, Obviously, I also go to my checkup twice a year every six months.

Renneth expressed her desire to manage herself well to avoid complications of SCD and the associated mortality. She explained that one of her ways of managing her SCD and avoiding VOC is by taking her prescribed hydroxyurea and avoiding stressful situations such as pregnancy. She said:

> And I am not planning to get pregnant. I'm not planning on carrying anything. I'm

> not dealing with any of that because my body can't take it. Because I know that I have to come off of the medications, my hydroxyurea, that's helping me stay out of the ER and out of the hospital. I was actually part of the case study of Hydroxyurea at the very beginning of it when I was 16 or 17 something like that. So I've been on it for quite some time and that has definitely been a huge lifesaver for me, because I was no longer in the ER every month, or in the hospital every month, and it's allowed me to have more of a life. But if I was going to get pregnant, I would have to come off of that. I would have to come off some of that other drugs that I take and then I would probably have more sickle cell crisis where I'm out of work more often, and I'm in the ER or the hospital more often. And it's just kind of like, I don't want to do that to myself and I don't want to do that to my child.

Madden Girl explained that she is able to manage herself as she knows and sets her limits daily so she can avoid vaso-occlusive crises. She states:

> So I would say there's a lot of restrictions, but you have to, you have to know yourself. You have to learn yourself. That's what sickle cell requires. It requires you to know yourself enough to know what you can do versus what you think you can do. You have to know the difference. At one point I was terrified to travel. I didn't want to go anywhere else and get sick and they have no idea about this illness and what to do. I wasn't doing what normal 19-year old, 20-year-old were doing at the time. Spring break comes. Everybody's somewhere else and I'm still here. And I had to take the time out and learn myself, learn my body, what can my body handle? What can I do that won't be too much and won't cause a disruption

> for anyone else? I took that time, I sat back. I'm like, okay, if I want to go somewhere I need to make sure my blood work is good, make sure my hemoglobin is high, make sure my white blood count is stable, and then start planning.

Jaz explained how she sometimes uses home remedy to manage her sickle cell disease. She said:

> So yeah, I got home and I can't feel my leg, and it really started freaking me out, and I went online to figure out what can I do about this. And I wished I knew that lady. They told me online that a lot of times the sickler, you can have a minor stroke and that could be a reason for that. Anyway, when I went to research further preventatives or what I can do to try to stop this, you know what popped up? Rosemary oil. I went to the vitamin shop, bought rosemary oil, and I kid you not. Three times a day, I was just rubbing on my leg. Gone, yeah. The feeling was gone. Honestly, I would do it all holistic, the yams and all the different kinds of foods and stuff to keep me healthy. Medications to me, I feel like they're designed to hurt me more. So and I'm figuring, okay, well our ancestors had this same problem many, many years ago and they did not have Demerol and all these different things to take. They dealt with herbs and stuff like that, and I tell you just doing it that way, I feel a whole lot better.

The narratives above portray how SCD individuals use self-care maintenance, self-care monitoring, and self-care management to minimize and suppress the frequency of their VOC and its complications. The themes that emerged from this study have been identified as *Excruciating Pain, Depressing, Feeling Helpless,* and *Dreading Healthcare*

Workers' Attitude. In what seems like a simple description, each participant verbalized that going through VOC as a SCD sufferer is very difficult and challenging. The results of this study revealed that there are physical and psychological challenges associated with VOC. Regardless of gender, age, occupation, and the number of VOCs per year, all participants expressed that they experience *Excruciating Pain, Depressing, Feeling Helpless,* and *Dreading Healthcare Workers' Attitude.*

The lyrics from the song of Prodigy (1974-2017) below (Prodigy, 2011) best captures the experience of sickle cell disease people as related to VOCVOC for individuals with SCD:

> Picture yourself standing at a bus stop on a particularly cold wintry morning in Long Island, New York.
>
> The wind coming across Lake Hempstead whips at your face as you burrow deeper into your jacket for warmth.
>
> Suddenly, a fiery pain slowly begins to creep into your ankles.
>
> Triggered by the frigid temperature, the pain begins working its way up through your legs then into your torso and arms.
>
> Before long, your entire skeleton feels as if it's on fire (My Infamous Life, 2011, Prodigy).

Significance of the Study

The significance of this qualitative phenomenological study was to explore the lived experience of adults with sickle cell disease during vaso-occlusive crises. Review of the literature provided a scarcity of studies that addressed the phenomena of interest.

Knowledge and understanding of the emerging themes of this study support the "how" and "what" of the experience of adults with sickle cell during VOC. Four overarching themes were identified as: *Excruciating Pain; Feeling Helpless; Depressing; Dreading Healthcare Workers' Attitude*. The data generated from this study adds to the body of phenomenological research and other disciplines such as education, psychology, social work, and medicine, as the care of sickle cell individuals is multidisciplinary. The data generated from this study adds to the body of nursing knowledge and advances nursing science.

Significance of the Study to Nursing

The findings from study have an impact on the current and future status of the nursing profession. Nursing research provides the scientific basis for the practice of nursing profession. This study increases the profession's body of scientific knowledge by filling a gap in the knowledge. There are specific implications for nursing education, nursing practice, nursing research, and health/public policy that will be discussed.

Implications for Nursing Education

The findings from this study could be incorporated into nursing curriculum. Participants verbalized that some healthcare providers had little or no knowledge about sickle hemoglobinopathies. This study shed light on the experience of SCD people during VOC that cannot be found in traditional nursing school textbooks. The findings from this study could be incorporated into a Fundamentals of Nursing course whereby nursing students will be aware of this genetic disease and treat individual with SCD with sensitivity. In addition, courses could be added to the Associate Nursing (ASN) and Bachelor of Science in Nursing (BSN) curricula highlighting different sickle

hemoglobinopathies. The findings from this study can help hospitals add SCD topics to their staff development courses to educate their nurses better on the sickle cell VOC.

Implications for Nursing Practice

The aim of this study was to give adults with SCD a voice to express their lived experience during VOC. The findings from this study highlights what it means to be in VOC and the complications that associated with it as experienced by the study participants. Within the findings, it is evident that individuals with sickle cell disease go through serious excruciating pain, some form of isolation, helplessness, and fear of negative attitudes of healthcare workers, especially doctors and nurses. Participants verbalized that they wish healthcare workers would understand what they go through during VOC. The findings of this study may help doctors and nurses understand the seriousness of the pain of VOC which may help improve the care of these patients during VOC. The findings from this study will help healthcare workers understand the incongruity between verbal and nonverbal cues from patients during VOC, and thus help eliminate the assumption that SCD patients are addicted and seeking pain medication, an assumption that may lead to their pain being left untreated properly. The findings from this study may help structure the practice of nursing such that more focus is on providing adequate nursing care to SCD patients during VOC without of preconceptions and condemnatory attitudes. This study will help to reassess and strengthen the cultural sensitivity training and education provided to nurses and other healthcare professionals. This study will help nurses understand that there is increased potential for prolonged admissions, hospitalizations, and readmissions when pain crises are mismanaged. This

study will improve nurses' approach to care delivery to patients with SCD so these patients can have better quality of life and avoid premature mortality.

Implications for Nursing Research

This phenomenological study explored the lived experience of adults with SCD. Understanding the "how" and the "what" of each participant' experience of VOC further enhances the knowledge of how to support this special group of people. A review of the literature reveals that there is a lack of studies that address the lived experience of sickle cell patients during vaso-occlusive crises. The findings from this study help fill those gaps. Therefore, this study adds to the body of knowledge on the meaning of being a sickle cell individual experiencing VOC. This study offers insight regarding to what SCD patients endure during VOC, both physically and psychologically. According to Reich, Cantrell and Smeltzer (2019), despite that multiple interventions can be used to treat and probably cure SCD, these interventions are used infrequently. They explained that this is related to provider's discomfort with the use of treatments options which contribute to poor quality of care and a subsequent increase in mortality in adults with sickle cell disease.

This study's findings could lead to more phenomenological studies being done in other chronic illnesses, genetic illnesses, and in different populations. The findings could also lead to replication of a study like this in other parts of the nation and globally. The findings from this study regarding the differences and similarities in the experiences of people suffering from chronic illnesses could possibly bring a national and or global change. The findings of this transcendental phenomenology study could contribute to scientific discussions about SCD patients' experience of VOC. The findings of this study

could inspire further investigation into ways to minimize the suffering of people affected by SCD, prevent the spread of the disease, and reduce the associated mortality rate. The findings from this study could bring more awareness of phenomenological studies and their contribution to nursing research.

Implications for Health/Public Policy

Currently, patient satisfaction determines many things including reimbursement for hospitalization. The findings of this study could point out the barriers to attaining complete patient satisfaction with every encounter. Participants voiced their opinion regarding their care in the emergency department that could possibly affect the patient satisfaction surveys. Healthcare organizations and nurses should develop policies that reinforce the ANA code of ethics and provide compassionate care for SCD patients. Bioethical principles such as respect, beneficence (doing good), and justice (treating people fairly) should be emphasized. The findings of this study could update practice and policy regarding continuous staff education to prepare health care professionals with new ways to teach SCD patients and their families on pain management and coping strategies, thus reducing the frequency VOC. This study can educate legislators improve SCD treatment, research, prevention, and monitoring. It can encourage the legislators to provide more benefits in the Medicaid program and make available federal funds for education and outreach to Medicaid eligible adults with SCD.

The findings of this study could inspire legislators and policy-makers to reinforce effective and adequate pain management. Law makers could develop ways for making sure all healthcare workers abide by pain management guidelines and take pain management more seriously. The findings of this study could provide lawmakers with

additional knowledge to inform them about the appropriate and recommended levels of care and services for people living with SCD. Furthermore, better understanding of the studied phenomenon among lawmakers could lead to the implementation of public educational trainings to increase the awareness of SCD. This study offers valuable information in order to propel health and public policy and possible regulatory changes in the pain management of individual with SCD during VOC.

Strengths and Limitations of the Study

Every study has strengths and limitations. This study's strengths included rich dialogue from 20 participants on the lived experience of being in vaso-occlusive crises as sickle cell individuals. The rich descriptions of the phenomena provide a better understanding of the essence of the experience. The sample utilized was diverse and represented participants with SCD who have experienced different numbers of VOCs per year. Moustakas' transcendental phenomenology method provided a systematic approach ideal for this novice researcher. The epoche process allowed the researcher to put aside preconceptions and personal biases in order to be open and receptive to discover the essence of individuals' experience.

The limitations of this study included the fact that participants drawn only in South Florida and only represented people of African descent, and there are other ethnic groups also at risk of SCD including Hispanics from Central and South America, Middle Easterners, etc. hence, limiting the transferability of the findings. While the sample was varied, only SCD individuals who were 18-65 met the inclusion criteria, the study did not capture the experience of children under age 17 years or persons over age 65 years. Other limitations included the researcher being a novice and Moustakas' approach requiring a

thorough understanding of the complex concepts of transcendental phenomenology. In addition, the inclusion criteria included only SCD people who were fluent in English, therefore, the lived experience of VOC in non-English speaking individuals was not captured.

Recommendations for Future Study

This study contributes to understanding what it is to experience VOC as a SCD person. It highlights physical and psychologic sufferings such as *Excruciating Pain, Depressing,* and *Feeling Helpless.* They also expressed their fear of stigmatization and *Dreading Healthcare Workers' Attitude.* More research on this group of individuals with SCD is needed to add to the body of nursing science. Additional qualitative studies should be conducted to investigate the strategies used by other ethnic group also at risk for SCD and VOC to cope with their physical and psychological suffering. This study should be replicated in other populations of individuals with SCD such as children and other people from other parts of the world. Correlational studies should examine the relationship between increased knowledge of SCD management and frequency of VOC. Correlational studies to determine the relationships among the themes identified in this study. In addition, studies that explore how SCD persons deal with their financial hardship should be done. Studies to examine if the pain management in ER in hospitals during VOC are compliant with the American Pain Association recommendations.

Conclusion

The lived experience of 20 SCD individuals during VOC was explored using Moustakas' (1994) transcendental phenomenology as a guide. Face-to-face interviews were utilized to collect data, which were then transcribed by a transcriptionist, reviewed

by the researcher, and authenticated by the participants. The data was analyzed by the researcher. Individual textural and structural descriptions were composed after each interview. Once data saturation was achieved, four themes: *Excruciating Pain, Depressing, Feeling Helpless,* and *Dreading Healthcare Workers' Attitude* emerged from the analysis. Participants were presented with the themes and confirmed the relevance. A composite of the textural and structural descriptions was synthesized to uncover the essence of the meaning of the participants' experience.

The participants described that being in vaso-occlusive crises as a SCD individual is physically and emotionally challenging. The results of this study found that regardless of the frequency of VOC, age or gender, VOC still pose physical and psychologic sufferings and challenges such as *Excruciating Pain, Depressing, Feeling Helpless, Dreading Healthcare Worker' Attitudes.* Despite these difficulties and challenges experienced by this group of people during VOC, sickle cell individuals continue to face inadequate pain management, ineffective disease management, and health disparity.

In the end, the participants expressed their desire for more research on SCD, more treatment options. They voiced out their concern about the need for better treatment and management of the disease. They further explained that increasing the knowledge of healthcare workers about the disease might improve their attitudes toward SCD patients especially in the ED. Moreover, these participants feel that SCD should be a significant part of nursing and medical school curricula. The review of the literature indicated that understanding SCD individuals' experience during VOC is important in order to adequately care and support them better during these critical times of their lives. The

contribution of this special group of people who were part of this study is priceless as they gave a voice to all sickle cell patients and their experience during this painful VOC.

References

Adenike, F. F., Matthew I. O., & Lawal, Y. R. (2018). Knowledge of sickle cell disease and pre-marital genotype screening among students of a tertiary educational institution in south western Nigeria. *International Journal of Caring Sciences,.11 (1), 286.*

Adeyemo, T. A., Ojewumi, O. O., Diaku-Akinwumi, I. N., Ayinde, O. C., & Alani, S. (2015). Health related quality of life and perception of stigmatization in adolescents living with sickle cell disease in Nigeria: a cross sectional study. *Pediatric Blood & Cancer, 62 (7), 1245-1251.*

Adis Medical Writers (2014). Both prevention and treatment are important when managing sickle cell disease. *Drugs & Therapy Perspectives, 30, 411–416.*

Adzika, V. A., Glozah, F. N., Ayim-Aboagye, D., & Ahorlu, C. K. (2017). Socio-demographic characteristics and psychosocial consequences of sickle cell disease: the case of patients in a public hospital in Ghana. *Journal of Health, Population and Nutrition 36 (4), 1-10.*

Altman, T. (2010). Attitude: A concept analysis. *Nursing Forum 43 (3), 144-50.*

Amara, J. L., Almeida, N. A., Santos, P. S., Oliveira, P. P., & Lanza, F. M. (2015). Socio-demographic, economic and health profile of adults with sickle-cell disease. *Rev Rene, 16 (3), 296-305.*

American Nurses Association (2015). *Code of ethics for nurses with interpretive statements.* Retrieved from http://www.nursingworld.org/codeofethics.

Audulv, A. (2013). The over time development of chronic illness self-management patterns: a longitudinal qualitative study. *BMC Public Health, 13 (452), 1-15.*

Badawy, S.M., Barrera, L., Cai, S., Thompson, A.A. (2018). Association between Participants' characteristics, patient-reported outcomes, and clinical outcomes in youth with sickle cell disease. *BioMed Research International, 1-8.*

Banks, M., & Shikle, J. (2018). Hyperhemolysis syndrome in patients with sickle cell disease. *Archives of Pathology & Laboratory Medicine, 142, 1425-1427.*

Berg, B. L., & Lune, H. (2012). *Qualitative research methods for the social sciences* (8th ed.). Boston, MA: Pearson Education, Inc.

Barriteau, M. & McNaull. M. (2018). Sickle cell disease in the emergency department: complications and management, *Clinical Pediatric Emergency Medicine,*19 (2), 103-109.

Berezow, A. (2018). White overdose deaths 50% higher than Blacks, 167% higher than Hispanics. *American Council of Science and Health.*

Bergman, E.J., & Diamond, N.J. (2013). Sickle cell disease and the "difficult patient" conundrum. *The American Journal of Bioethics, 13(4): 3–10.*

Boyle, S. M., Jacobs, B., Sayani, F. A., Hoffman, B. (2016). Management of the dialysis with sickle cell disease. *Seminars in Dialysis, 29 (1), 62-70.*

Bundy, D. J., Richardson, T. E., Hall, M., Raphael, J. L., Brousseau, D. C., Arnold, S. D., Kalpatthi, R. V., Ellison, A. V., Oyeku, S. O., Shah, S. S. (2017). Association of guideline-adherent antibiotic treatment with readmission of children with sickle cell disease hospitalized with acute chest syndrome. *JAMA Pediatrics, 171 (11), 1091-1099.*

Cacciotti, C., Vaiselbuh, S., & Romanos-Sirakis, E., (2017). pain management for sickle cell disease in the pediatric emergency department: medications and hospitalization trends. *Clinical Pediatrics, 56(12,) 1109–1114.*

Carroll, L.J. (2015). Perceived pain management satisfaction in adult patients with sickle cell disease experiencing a vaso-occlusive crisis. *The Journal of Chi Eta Phi Sorority, 59(1).*

Carter, D., Sendziuk, P., Eliott, J.A., & Braunack-Mayer, A. (2016). Why is pain still under-treated in the emergency department? two new hypotheses. *Bioethics, 30 (3), 195–202.*

Carlton Haywood, Tanabe, P, Naik, R., Beach, M. C., & Lanzkron, S. (2013). The impact of race and disease on sickle cell patient wait times in the emergency department. *American Journal of Emergency Medicine, 31 (4), 651-656.*

Carvalho, E. M., Santos, F. M., Izidoro, C., Caldeira dos Santos. M. L., Batista Santos, R. (2016). Nursing care to people suffering with sickle cell disease in emergency unit. *Ciencia Cuidado Saude, 15 (2), 328-335.*

Cevher, F., Unal, S., Kucukkavruk, S., Tasdelen, B., & Tunctan, B. (2018) Quality of life, clinical effectiveness, and satisfaction in pediatric and young adult patients with sickle cell disease receiving hydroxyurea therapy. *Turkiye Klinikleri Journal of Medical Sciences, 38 (1), 30-39.*

Coleman, B., Ellis-Caird, H., McGowan, J., & Benjamin, M. J. (2016). How sickle cell disease patients experience, understand and explain their pain: an interpretative phenomenological analysis study. *British Journal of Health Psychology, 21, 190–*

203.

Converse, M. (2012). Philosophy of phenomenology: how understanding aids research. *Nurse Researcher, 20 (1), 28-32*

Costa, D. O., Araújo, F. A., Araújo, L. S., Batista da Silva, U., Santos, E. A., Ferreira, S. L., & Xavier, A. S. (2018). Self-care of men with priapism and sickle cell disease. *Revista Brasileira de Enfermagem, 71 (5), 2418-24.*

Creswell, J. W., & Poth, C.N. (2017). Qualitative inquiry and research design: Choosing among five approaches (3rd ed.). Thousand Oaks, CA: Sage.

Creswell, J. W. (2013). Qualitative inquiry and research design: Choosing among five approaches (3rd ed.). Thousand Oaks, CA: Sage.

Crosby, E.L., Joffe, L.E., Reynolds, N., Peugh, J.L., Manegold, E., & Ahna L. H. (2015). Psychometric properties of the psychosocial assessment tool-general in adolescents and young adults with sickle cell disease. *Journal of Pediatric Psychology, 41(4), 2016, 397–405.*

Crotty, M. (2013). The foundations of social research: Meaning and perspective in the research process. Thousand Oakes, CA: Sage.

Crowther, S., Ironside, P., Spence, D., & Smythe. L. (2017). Crafting stories in hermeneutic phenomenology research: a methodological device. *Qualitative Health Research, 27(6), 826–835.*

Cutler, S., Crawford, P., & Engleking, R. (2018). Effectiveness of group self-management interventions for persons with chronic conditions: a systematic

review. *Medsurg Nursing, 27 (6), 359-367.*

Cunningham, T, J., Croft, J. B., Liu, Y., Lu, H., Eke, P.K., & Giles, W. H. (2017). Vital signs: racial disparities in age-specific mortality among Blacks or African Americans — United States, 1999–2015. *Morbidity and Mortality Weekly Report, 66 (17).*

Damille Silva, H., Paixão, G.P., Santos Silva, C., Bittencour, I. S., Evangelista, T.J., & Souza da Silva, R. (2014). Sickle cell disease and its psychosocial aspects: the look of patient and caregiver family. *Revista Cuidarte, 475-483.*

Darbari, D. (2018) Opioid-crisis-affects-sickle-cell-patients-presenting-with-related-pain Opioid. *Retrieved form www.ahcmedia.com/articles/141851*

Davis, M., & Brown, T. L. (2016). Major depressive disorder and sickle cell anemia: bidirectional comorbidities. *Hematology & Transfusion International Journal, 2 (3).*

Desai, G., Shah, P., Dave, K., Bhatt, H., Shrey Desai, Modi, D., Anand, A., & Shah, S (2017). Sickle cell disease and pregnancy outcomes: a study of the community-based hospital in a tribal block of Gujarat, India. *Journal of Health, Population and Nutrition (36) 3, 1-7.*

Elahi, N, Tahery, N., Ahmadi, F., Rostami, S., & Elahi, N. (2017) Lived experienced of patients with sickle cell disease anemia about disease management (a qualitative research). *Annals of Tropical Medicine and Public Health, 10(5), 1238-1242.*

Ellis, P. (2016). The language of research (part 8): phenomenological research. *Wounds UK, 12 (1), 128-129.*

Elsbach, K. D., & Kramer, R. M. (2015). *Handbook of qualitative organizational research: innovative pathways and methods.* New York, NY.

Ezenwa, M. O., Yao, Y., Wilkie, D.J., Molokie, R. E., Wang, Z. J., Mandernach, M. W., & Suarez, M. L. (2017). Coping with pain in the face of healthcare injustice in patients

with sickle cell disease. *Journal of Immigrant Minority Health, 19, 1449–1456.*

Faremi, A.F., Olatubi, M. I., & Lawal, Y. R. (2018). Knowledge of sickle cell disease and pre-marital genotype screening among students of a tertiary educational institution in south western Nigeria. *International Journal of Caring Sciences, 11 (1), 285*.

Felix de Freitas, S.L., Ivo, M. L., Figueiredo, M.S., Souza Gerk, M. A., Nunes, C. B., & Monteiro, F. F. (2018). Quality of life in adults with sickle cell disease: an integrative review of the literature. *Revista Brasileira de Enfermagem, 71(1):195-205.*

Franceschi, L. D., Mura, P., Schweiger, V., Vencato, E., Quaglia, F. M., Delmonte, L., Evangelista, M., Polati, E., Olivieri, O., & Finco, G. (2016). Fentanyl buccal tablet: a new breakthrough pain medication in early management of severe vaso-occlusive crisis in sickle cell disease. *Pain Practice, 16, (6), 680–687.*

Fosdal, M. B. (2015). Perception of pain among pediatric patients with sickle cell pain crisis. *Journal of Pediatric Oncology, 32 (1), 5-20.*

Gardner, C., Boll, D., Bhosale, P., & Jaffe, T.A. (2016). CT abdominal imaging findings in patients with sickle cell disease: acute vaso-occlusive crisis, complications, and chronic sequelae. *Abdominal Radiology, 41:2524–2532.*

Gesteira, R, Bousso, S., Misko, R.D., Ichikawa, F.M., & Oliveira, P. Families of children with sickle cell: an integrated review. *Online Brazilian Journal of Nursing, 15(2), 276-290.*

Gomes. L. M., Torres, H. C., Viana, M. B., Pereira. I. A., & Caldeira, A. P. (2014). Access and care of individuals with sickle cell anemia in a primary care service. *Acta Paul Enferm, 27 (4), 348-55.*

Graves, J. K., Hodge, C., & Jacob, E. (2016). Depression, anxiety, and quality of life in children and adolescents with sickle cell disease. *Pediatric Nursing, 42 (3), 113-144.*

Hamm, J., Rathore, N., LeBlanc, Z., Lebensburger, J., Lee. P., Kwiatkowski, J. L., & Meier, E. M. (2016). Cranial epidural hematomas: A case series and literature review of this rare complication associated with sickle cell disease. *Pediatric Blood & Cancer, 64, 1-8.*

Harrison, S., Walcott, C. M., & Warne, T .D. (2016). Knowledge and awareness of sickle cell trait among young African American adults. *Western Journal of Nursing Research,* 1222-1239.

Hassell, K.L. (2010). Population estimates of sickle cell disease in the U.S. *American Journal of Preventive Medicine, 38 (4), 512-521*.

Haywood, C., Lanzkron, S., Bediako, S., Strouss, J. J., Haythornthwaite, J., Carroll, C. P., Diener-West, M., Onojobi, G., & Beach, M. C. (2014). Perceived discrimination, patient trust, and adherence to medical recommendations among persons with sickle cell disease. *Journal of General Internal Medicine, 29(12), 1657–1662.*

Hulihan, M., Hassell, K. L., Raphael, J. L., Smith-Whitley, K., & Thorpe, P. (2017). CDC grand rounds: improving the lives of persons with sickle cell disease. *Morbidity & Mortality Weekly Report, 66 (46), 1269-1271.*

International Association for the Study of Pain. (2015). Pain from sickle cell disease prompts discussion at U.S. meeting. Retrieved from http://www.iasp-pain.org.

Jain, S., Bakshi, N., & Krishnamurti, L. (2017). Acute Chest Syndrome in Children with Sickle Cell Disease. *Pediatric Allergy, Immunology & Pulmonology, 30 (4), 191-201.*

Jenerette, C. M., Brewer, C. A., & Ataga, K. I. (2014). Care seeking for pain in young adults with sickle cell disease. *Pain Management Nursing, 15(1), 324-330.*

Jenerette, C. M., Pierre-Louis, B. J., Matthie, N., Girardeau, Y. (2015). Nurses' attitudes toward patients with sickle cell disease: a worksite comparison. *Pain Management Nursing, 16(3), 173-181.*

Jokelainen, J., Timonen, M., keinanen-Kiukaanniemi, Harkonen, P., Jurvelin, H., & Suija, K. (2019). Validation of the Zung self-rating depression scale (SDS) in older adults. *Scandinavian Journal of Primary Health Care 37 (3), 353–357.*

Keane, B & Defoe, L. (2016). Supported or stigmatized? The impact of sickle cell disease on families. *Community Practitioner, 44-47.*

Kim, S., Brathwaite, R. & Kim, Ook (2017). Evidence-based practice standard care for acute pain management in adults with sickle cell disease in an urgent care center. *Quality Management in Health Care, 26 (2), 108–115.*

Kuhn, T. S. (2012). *The structure of scientific revolutions* (4th ed.). University of Chicago Press, Chicago.

Lakkakula, B. V., Sahoo, R., Verma, H., Lakkakula, Saikrishna (2018). Pain management issues as part of the comprehensive care of patients with sickle cell disease. *Pain Management Nursing, 19 (6), 1-15*.

Lazio, M. P., Costello, H. H., Courtney, D. M., Martinovich, D., Myers, R., Zosel, A., & Tanabe, P. (2010). A comparison of analgesic management for emergency department patients with sickle cell disease and renal colic. *The Clinical Journal of Pain, 26 (3), 199-205.*

Lee, M. L., & Mark, L. (2014). When a blood donor has sickle cell trait: Incidental findings and public health. *Hasting Center Report, 44*(3).

Lentz, M. B., & Kautz, D. D. (2017). Acute vaso-occlusive crisis in patients with sickle cell disease. *Nursing, 47 (1), 67-68.*

Lima-Filho, N. N., Campos, O., Figueiredo, M. S., Vicari, P., Cancado, R., Camargo Carvalho, A. C., & Bordin, J. O. (2016). Exercise-induced abnormal increase of systolic pulmonary artery pressure in adult patients with sickle cell anemia: an exercise stress echocardiography study. *Echocardiography, 33 (12), 1880-1890.*

Lincoln, Y. S., & Guba, E. G. (1985). *Naturalistic inquiry.* Newbury Park, CA: Sage.

Lincoln, Y. S. & Guba, E. G., (1994). Competing paradigms in qualitative research. *Handbook of Qualitative Research*. Thousand Oaks, CA: Sage.

Ludwig, N. N., Sil, S., Khowaja, M. K., Cohen, L. L., & Dampier, C. (2018). Executive functioning mediates the relationship between pain coping and quality of life in youth with sickle cell disease. *Journal of Pediatric Psychology, 43 (10), 1160-1169.*

Madubata, I. J., Odafe, M. O., Talavera, D. C., Hong, J. H., & Walker, R. L. (2018). Helplessness mediates racial discrimination and depression for African American young adults. *Journal of Black Psychology, 44(7), 626–643.*

Mainous III, A. G., Tanner, R. J., Harle, C. A., Baker, R., Shokar, N. K. & Hulihan, M. M. (2015). Attitudes toward management of sickle cell disease and its complications: a national survey of academic family physicians. *Anemia, vol. 2015, Article ID 853835, 6 pages.*

Mammen, C., Matsell, D., & Bissonnette, M. (2017). Acute kidney injury in children with sickle cell disease - compounding a chronic problem. *Pediatric Nephriology, 32 (98), 1287-1291.*

Manwani, D., & Frenette, P. S. (2013). Vaso-occlusion in sickle cell disease: pathophysiology and novel targeted therapies. *Blood, 122(24): 3892–3898.*

Matthie, N. & Jenerette, C. (2015). Sickle cell disease in adults: developing an appropriate care plan. *Clinical Journal of Oncology Nursing, 19 (5), 562-568.*

Matua, G.A. (2015). Choosing phenomenology as a guiding philosophy for nursing research. *Nurse Researcher, 22 (4).*

Mburu, J., & Odame, I. (2019). Sickle cell disease: reducing the global disease burden. International Journal of Laboratory Hematology, 1 (41), 82-88.

McBain, H., Newman, S., & Shipley, M. (2015). The impact of self-monitoring in chronic illness on healthcare utilization: a systematic review of reviews. *Biomed Central Health Services Research, 15 (565), 1-10.*

Miller-Matero, L. R., Chipungu, K., Martinez, S., Eshelman, A., & Eisenstein, D. (2017). How do I cope with pain? Let me count the ways: awareness of pain coping behaviors and relationships with depression and anxiety. *Psychology, Health & Medicine, 22 (1), 19-27.*

Morgan, K.A., Scott, J., Parshad-Asnani, M., Gibson, R. C., Garo, K. N., Lowe, G. A., Edwards, D., Abel, W. D., Reid, M., Haye, W, D., & Edwards, Christopher (2014). Associations amongst disease severity, religious coping and depression in a cohort of Jamaicans with sickle-cell disease. *Mental Health, Religion & Culture, 17 (9), 937-945.*

Moustakas, C. (2012). *Phenomenological research methods*. Thousand Oaks, CA: Sage.

Moustakas, C. (1994). *Phenomenological research methods*. Thousand Oaks, CA: Sage.

Moustakas, C. (1990). *Heuristic research: design, methodology, and applications.* Thousand Oaks, CA: Sage.

Munhall, P. (Ed.). (2012). *Nursing research: A qualitative perspective* (5th ed.). Sudbury, MA: Jones & Barlett Learning.

Myers, M., & Eckes, E. J. (2012). A novel approach to pain management in persons with sickle cell disease. *Medsurg Nursing, 21 (5), 293-298.*

Nansseu, J. N., Yanda, A. N., Chelo, D., Tatah, S. A., Awa, H. D., Seungue, J., & Koki, O. N. (2015). The Acute Chest Syndrome in Cameroonian children living with sickle cell disease. *BioMed Central Pediatrics, 15 (131), 1-8.*

National Heart, Lung, and Blood Institute, 2014. Sickle cell disease. Retrieved from http://www.nhlbi.nih.gov.

National Institute of Health, 2018. Sickle cell anemia. Retrieved from http://www.nih.gov.

Nelson, S. C., & Hackman, H. W. (2013). Race matters: perceptions of race and racism in a sickle cell center. Pediatric Blood Cancer, 60, 451–454.

Nottage, K.A., Hankins, J.S., James, D.M., Wang, W. C., Cancio, M.I., Faughnan, L.G., Richardson, J., Christensen, R., Guolian, K., Smeltzer, M & Anghelescu, L. (2016). Addressing challenges of clinical trials in acute pain: the pain management of vaso- occlusive crisis in children and young adults with sickle cell disease study. *Clinical Trials 2016, 13(4) 409–416.*

O'Connor, S., Hanes, D., Lindsey, A., Weiss, M., Petty, L., & Overcash, J. (2014). Attitudes among healthcare providers and patients diagnosed with sickle cell disease: frequent hospitalizations and stressors. *Clinical Journal of Oncology Nursing, 18 (6).*

Pizzo,E., Laverty, A.A., Phekoo, K.J., AlJuburi, G., Green,S.A., Bell, B., Majeed (2014). A retrospective analysis of the cost of hospitalizations for sickle cell disease with crisis in England, 2010/11. *Journal of Public Health |37 (3), 529 – 539.*

Polit, D. F., & Beck, C. T. (2017). *Nursing research: Generating and assessing evidence for nursing practice* (10th ed.). Philadelphia, PA: Lippincott, Williams & Wilkins.

Polizzi, D. (2019). The impossibility of criminal justice ethics: toward a phenomenology of the possible. *International Journal of Offender Therapy and Comparative Criminology, 63 (1), 135–153.*

Raji, S. O., Lawani, A. O., & James, B. O. (2016). Prevalence and correlates of major depression among Nigerian adults with sickle cell disease. *International Journal of Psychiatry Medicine, 51 (5), 456-466.*

Reich, M., Cantrell, A., Smeltzer, S. (2019). Sickle cell disease in the young adult population: a historical review and implications for nurses. *Medsurg Nursing, 24 (4), 213-218.*

Relling, J. (2014). Peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia. *Journal of the American Medical Association, 312 (10).*

Rich, S., Graham, M., Taket, A., Shelly, J. (2013). Navigating the terrain of lived experience: the value of lifeworld existentials for reflective analysis. *International Journal of Qualitative Method, 12, 498-510.*

Riegel, B., Jaarsma, T. & Strömberg, A. (2012), A middle-range theory of self-care of chronic illness. *Advances in Nursing Science, (35), 3, 194-204.*

Rivolo, M. (2018). Sickle as a holistic treatment approach to sickle cell disease related ulcers. *British Journal of Nursing, 27 (20), 6-10.*

Rooij, V. N. (2018). The societal impact of pain. *Pain Practice, 18, 6-142.*

Saganuwan, A. (2016). The pattern of sickle cell disease in sickle cell patients from northwestern Nigeria. *Clinical Medicine Insights: Therapeutics, 54-57.*

Santos, J., Jones, S., Wakefield, D., Grady, J., & Andemariam, B. (2016). Patient controlled analgesia for adults with sickle cell disease awaiting admission from the emergency department. *Pain Research & Management, 5, 1-5.*

Savitt, T. L., Wally, R., Haywood, C., & Creary, M. S. (2014). Use of the word "crisis" in sickle cell disease: the language of sickle cell. *Journal of the National Medical Association, 106 (1)23-30.*

Shardonofsky, J., Cesario, S. K., Fredland, N., Landrum, P., Hiatt, P. W., & Shardonofsky, Felix (2019). The lived experience of fathers caring for a child with cystic fibrosis. *Pediatric Nursing, 45 (2), 87-92.*

Simsek, N., Evli, M., and Uzdil, N., (2019). Learned resourcefulness, anxiety and depression levels in patients with chronic Pain. *International Journal of Caring Sciences, 12 (1), 240-247.*

Smith, W. R. (2014). Treating pain in sickle cell disease with opioids: clinical advances, ethical pitfalls. *Journal of Law, Medicine & Ethics, 42 (2), 139-146.*

Tembo, A. (2016). Phenomenological inquiry as a methodology for investigating the lived experience of being critically ill in intensive care. *Journal of Intensive and Critical Care, 2 (1:8), 1-5.*

Thomas, T. A., & Simmons, M. (2017). Pharmacologic management of asthma and acute chest syndrome in patients with sickle cell disease. *Journal of the National Society of Allied Health, 29-35.*

Tolba, O. A., El-Shanshory, M. R., El-Gamasy, M. A., & El-Shehaby, W. A. (2017). Speckle tracking evaluation of right ventricular functions in children with sickle cell disease. *Annals of Pediatric Cardiology, 10 (3), 230-233.*

Van Manen, M. (2014). *Phenomenology of practice: Meaning giving methods in phenomenological research and writing.* Walnut Creek, CA: Left Coast Press

Wakefielda, E. O., Zempskya, W. T., Puhl, R. M., & Litt, M. D. (2018). Conceptualizing pain-related stigma in adolescent chronic pain: a literature review and preliminary focus group findings. *Innovations in Pediatric Pain Research and Care, e679.*

Walker, C. A. (2015). Social constructionism and qualitative research. *Journal of Theory Construction & Testing, 19 (2), 37-38.*

Wonkam, A., Mba, Z. C., Mbanya, D., Ngogang, J., Ramesar, R., & Angwafo III, F. F. (2014). psychosocial burden of sickle cell disease on parents with an affected child in Cameroon. *Journal of Genetic Counseling, 23, 192–201.*

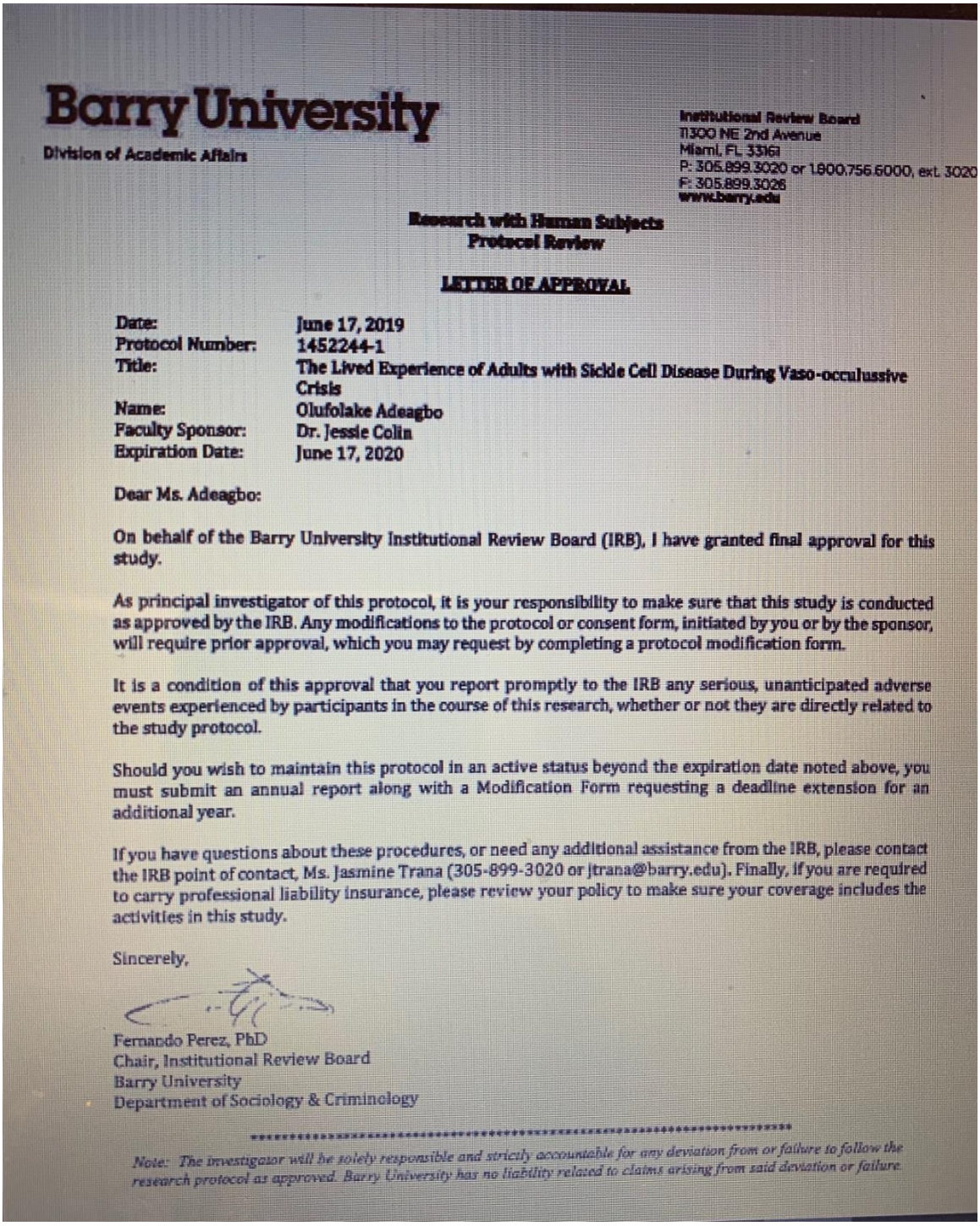
World Health Organization (2015).

Works, T., Jones, S., Grandy, J., & Andemariam, B. (2016). Traumatic exposure history as a risk factor for chronic pain in adult patients with sickle cell disease. *Health and Social Work, 41 (1).*

Xavier de Moraes, L., Bushatsky, M., Campos Barros, M., Ramos Barro, B., & Alves Bezerra, M.G (2017). Sickle cell disease: perspectives on the assistance provided in primary attention. *Fundamental Care Online, 9(3), 768-775.*

APPENDIX A

IRB APPROVAL LETTER



Barry University

Division of Academic Affairs

Institutional Review Board
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Miami, FL 33161
P: 305.899.3020 or 1.800.756.6000, ext. 3020
F: 305.899.3026
www.barry.edu

Research with Human Subjects
Protocol Review

LETTER OF APPROVAL

Date:	**June 17, 2019**
Protocol Number:	**1452244-1**
Title:	**The Lived Experience of Adults with Sickle Cell Disease During Vaso-occulussive Crisis**
Name:	**Olufolake Adeagbo**
Faculty Sponsor:	**Dr. Jessie Colin**
Expiration Date:	**June 17, 2020**

Dear Ms. Adeagbo:

On behalf of the Barry University Institutional Review Board (IRB), I have granted final approval for this study.

As principal investigator of this protocol, it is your responsibility to make sure that this study is conducted as approved by the IRB. Any modifications to the protocol or consent form, initiated by you or by the sponsor, will require prior approval, which you may request by completing a protocol modification form.

It is a condition of this approval that you report promptly to the IRB any serious, unanticipated adverse events experienced by participants in the course of this research, whether or not they are directly related to the study protocol.

Should you wish to maintain this protocol in an active status beyond the expiration date noted above, you must submit an annual report along with a Modification Form requesting a deadline extension for an additional year.

If you have questions about these procedures, or need any additional assistance from the IRB, please contact the IRB point of contact, Ms. Jasmine Trana (305-899-3020 or jtrana@barry.edu). Finally, if you are required to carry professional liability insurance, please review your policy to make sure your coverage includes the activities in this study.

Sincerely,

Fernando Perez, PhD
Chair, Institutional Review Board
Barry University
Department of Sociology & Criminology

****

Note: The investigator will be solely responsible and strictly accountable for any deviation from or failure to follow the research protocol as approved. Barry University has no liability related to claims arising from said deviation or failure.

APPENDIX B

INFORMED CONSENT FORM

Barry University

Informed Consent Form

Dear Research Participant:

Your participation in a research project is requested. The title of the study is The Lived Experience of Adults with Sickle Cell Disease During Vaso-occlusive Crises. The research is being conducted by Olufolake Adeagbo, a student in the Nursing department at Barry University, and is seeking information that will be useful in the field of Nursing. The aims of the research are to give this group of patients a voice to express their individual experience during vaso-occlusive crises, to provide an inductive description of the phenomenon and to gain understanding of the essence of the experience of having vaso-occlusive cries. In accordance with these aims, the following procedures will be used: individual interview. We anticipate the number of participants to be 30.

The participants will be informed about the purpose of the study, the meaning and essence of informed consent, the risks and benefits, as well as the process for recording the interview. They will be informed that there are no known benefits or risks for participating in the study. The participants will be advised that strict confidentiality will be maintained for the audio recordings and any notes that may be taken during the interview. Furthermore, a transcriptionist, who will be asked to sign a third-party agreement, will be transcribing digital audio recordings. The participants will be told that they can stop the interview at any time, and they can also refuse to answer a question(s) or ask that the recorder be turned off or suspended at any time during the interview. At the end of the interview, the researcher will thank the participant and remind him or her that privacy by the researcher will be maintained. The participants will be informed that data will be stored, transcribed, and used for the study. The audio recordings will be destroyed after the initial member check is completed.

If you decide to participate in this research, you will be asked to do the following: (1) fill out a demographic form lasting no longer than 10 minutes, (2) 60-minute interview face-to-face and (3) a 20-minute member check sessions over the telephone. The purpose of the member check session is for clarification and verification of the transcription of the initial interview. The total time for your participation in this study is 90 minutes.

If you decide to participate in this study, you will receive a $25 American Express gift card as a token of appreciation regardless if you complete the interview or not. Your consent to be a research participant is strictly voluntary and should you decline to participate, or should you choose to drop out at any time during the study, there will be no adverse effects on your health care. There are no known risks to you. Although there

are no direct benefits to you, your participation in this study may help our understanding of vaso-occlusive crises.

As a research participant, information you provide will be kept anonymous. No names or other identifiers will be collected on any of the instruments used. Any published results of the research will be in aggregate form and pseudonyms will be used in the study. All data will be kept in a locked file in a password protected personal computer in the researcher's office. A transcriptionist who has signed a third party confidentiality form will transcribe the audio recordings. Audio recordings will be destroyed after the member check session and transcripts of the recordings will be retained. Your signed consent form will be kept separate from the data. All data will be retained for 5 years upon completion of the study and indefinitely thereafter.

If you have any questions or concerns regarding the study or your participation in the study, you may contact me, Olufolake Adeagbo (Fola) at (954) 670-3769 or oadeagbo@mymail.barry.edu; my supervisor, Dr. Jessie M Colin at (305) 899-3800 or jcolin@barry.edu; or the Institutional Review Board point of contact, Jasmine Trana, at (305) 899-3020 or jtrana@barry.edu. If you are satisfied with the information provided and are willing to participate in this research, please signify your consent by signing this consent form.

Voluntary Consent

I acknowledge that I have been informed of the nature and purposes of this experiment by Olufolake Adeagbo and that I have read and understand the information presented above, and that I have received a copy of this form for my records. I give my voluntary consent to participate in this experiment.

_____	_____
Signature of Participant	*Date*

_____	_____	_____	_____
Researcher	*Date*	*Witness*	*Date*

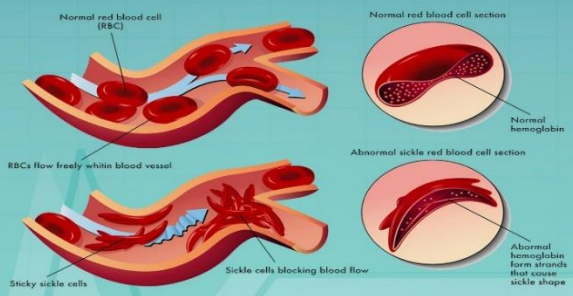
(Witness signature is required only if research involves pregnant women, children, other vulnerable populations, or if more than minimal risk is present.)

APPENDIX C

FLYER

SICKLE CELL DISEASE PATIENTS NEEDED

To participate in a research about sickle cell disease.



The purpose of the study is to discuss the experience of Sickle cell patients during pain crises

THE PARTICIPANTS MUST BE:

- An adult with sickle cell disease who has experienced at least a pain crisis
- Fluent in English

Available to complete:

- A Demographic form lasting no longer than 10 minutes
- A 60-minute face-to-face interview
- A 20 minute phone interview to confirm the transcribed interview

Total of 90 minutes for both interviews

INTERESTED?

For more information, contact the researcher listed below: (Fola)

Sponsor: Olufolake Adeagbo (Fola) Phone: 954 670 3769
Email: olufolake.adeagbo@mymail.barry.edu
University Faculty Sponsor: Jessie Colin, PhD, RN, FAAN
Phone: 305 899 3830 Email: jcolin@barry.edu
Institutional Review Board:Jasmin Trana

All participants will receive a $25 American Express gift card for their time

APPENDIX D

INTERVIEW QUESTIONS

1. What does it feel like to have sickle cell disease?
2. How many times do you have pain crises in a year?
3. Tell me how you feel during pain crises?
4. How many times do you visit the emergency room per year?
5. How would you describe the care that you receive during your pain crisis?
6. Please describe your needs during a crisis.
7. How do you think your lifestyle is affected by sickle cell disease?

APPENDIX E

DEMOGRAPHIC FORM

1. **Pseudonym: _____**

2. **Age:**

   □□18-30 □□41-50 □□60-65

   □□31-40 □□51-60

3. **Gender:**

   □ Female □□Male

4. **What is your race?**

   □□Black or African American □□White

   □□Hispanic or Latino □□ Asian

   □ Native Hawaiian or other Pacific Islander. □□American Indian or Alaska Native.

5. **What type of occupation do you have?**

   □□Professional □□Non-professional □

6. **What is your level of education?**

   □□Doctorate degree □□Bachelor's degree

   □□Master's degree □□Associate degree □□High School

7. **How many times do you have crises in a year?**

   □□1-2 □□3-4 □□5-6

   □ □□7-8 □□9-10

APPENDIX F

THIRD PARTY CONFIDENTIALITY FORM

Confidentiality Agreement for use with Transcription Services

Research Study Title: The lived experience of adults with sickle cell disease during vaso-occlusive crises.

1. I, _____ transcriptionist, agree to maintain full confidentiality of all research data received from the research team related to this research study.

2. I will hold in strictest confidence the identity of any individual that may be revealed during the transcription of interviews or in any associated documents.

3. I will not make copies of any audio-recordings or other research data, unless specifically requested to do so by the researcher.

4. I will not provide the research data to any third parties without the client's consent.

5. I will store all study-related data in a safe, secure location as long as they are in my possession. All audio recordings will be stored in an encrypted format.

6. All data provided or created for purposes of this agreement, including any back-up records, will be returned to the research team or permanently deleted. When I have received confirmation that the transcription work I performed has been satisfactorily completed, any of the research data that remains with me will be returned to the research team or destroyed, pursuant to the instructions of the research team.

Transcriber’s name (printed) _____

Transcriber's signature _____

Date _____

APPENDIX G

VITA

ACADEMIC DEGREES	DATES	DEGREE	MAJOR
Barry University	2016-2019	PhD	Nursing
Florida Atlantic University	2006-2009	MSN	Nursing
Florida Atlantic University	2003-2005	BSN	Nursing
Broward College	2002-2003	Non-Degree	Health Sciences
University College Hospital	1990-1993	Diploma	Nursing

PROFESSIONAL EXPERIENCE

Broward College	2012-present	Assistant Professor
Broward College	2009-2012	Adjunct Professor
Holy Cross Hospital	2000-2012	Nurse/ Asst. Manager
Holy Cross Hospital	2012-2016	PRN Charge Nurse

CERTIFICATION/LICENSURE

Advanced Cardiac Life Support (ACLS)

Basic Life Support (BLS)

Florida Board of Nursing Registered Licensure