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Priapism Among Male Adolescents with Sickle Cell Disease

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UNIVERSITY OF SAN DIEGO
Hahn School of Nursing and Health Science
DOCTOR OF PHILOSOPHY IN NURSING

PRIAPISM AMONG MALE ADOLESCENTS WITH SICKLE CELL DISEASE

by

Prince Ocansey

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requirements for the degree
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ABSTRACT

The purpose of this qualitative phenomenological study was to describe the lived experience of priapism among male adolescents with sickle cell disease (SCD). A painful genetic disorder with no cure, SCD severely impacts the lives of patients, as well as their families and friends. Priapism is one of the complications of SCD and involves persistent and painful penile erections. The review of literature established a gap in researchers' understanding of the lived experience of priapism among male adolescents with SCD. This study aimed to bridge that gap.

Using a convenience sampling method, 7 adolescent males between the ages of 16 and 19 with a history of SCD and the complication of priapism were selected for the study. A phenomenological approach guided this study. Qualitative data was drawn from semistructured face-to-face interviews using an interview protocol with 22 open-ended questions. Data were analyzed by transcribing audio tapes and coding information from transcripts into themes, patterns, and categories. Themes were identified that reflected the central, recurrent ideas that emerged from the participant responses.

The results of this research study demonstrate the significance of various themes associated with living with SCD and the complication of priapism for male adolescents, including: (a) Concerns with sexual performance, stigma, and associated psychological effects; (b) Quality of life and support systems; (c) Experiences with medical care; and (d) Knowledge of the condition and advice for others. The findings of this study confirm some previous literature while lending further insights that help expand researchers' knowledge regarding the lived experiences of male adolescents with SCD and the complication of priapism.

Based on these findings, the implications for practice and opportunities for future research are broad and varied. It is imperative to develop awareness trainings or educational programs to better inform medical practitioners and SCD patients about the complication of priapism. Further research is needed to gain a better understanding of the prevalence of priapism in the SCD male adolescent population. Future studies should analyze whether this complication is hereditary, lifestyle-related, or can affect anyone.

Keywords: priapism, male adolescent, sickle cell disease

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DEDICATION

I dedicate this dissertation to my wife, Elizabeth, and my children, Perry, Karen, Daniel, Brian, and Briana. Their steadfast encouragement, motivation, and support have been magnificent throughout this journey.

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CHAPTER 1

INTRODUCTION

Purpose of the Study

Sickle cell disease (SCD) is among the most frequently occurring inherited disorders in the United States. It affects an estimated 100,000 residents and occurs in one in every 500 African-American births on average (Powars, Chan, Hiti, Ramicone, & Johnson, 2005). A severe and painful genetic disorder with no cure, SCD severely impacts the lives of the patients as well as those of their families and friends. Priapism, one of the negative side effects of SCD involving a persistent and painful erection of the penis (Chrouser, Ajiboye, Oyetunji, & Chang, 2011), is a distressing complication that affects the quality of life of many males with SCD, especially adolescents.

The purpose of this study was to use the lived experience of priapism, via a qualitative phenomenological research design, to understand priapism among male adolescents with SCD. The use of participants' lived experiences aimed to provide an in-depth understanding of the lives of individuals with SCD, focusing on their individual perceptions of priapism. The research aims of this study are given in the following section.

Research Aims

The research aims of this study were as follows:

- Describe the effects of priapism on the self-esteem of male adolescents with SCD.
- Describe the impact of priapism on the social development of male adolescents with SCD.
- Explore the coping strategies used by male adolescents with SCD and priapism.

Statement of the Problem

A source of chronic pain, SCD is a health issue prevalent in many populations and is one of the leading genetic diseases throughout the world (Lauderdale, 2003). Recent developments in medical management and technology have helped patients with this disease live longer and better lives. Research has indicated that these advances have enabled most children living with this chronic illness to survive to adulthood (Erskine, 2011). Initiatives by health care providers aim to ensure that young people and adolescents living with SCD have access to continuing developments in treatment. As part of such initiatives, this research study was designed to understand the lived experiences of individuals who struggle with SCD-related priapism.

Most people are not aware of the symptoms of SCD nor possible ways to live with it. One complication of SCD is priapism, which is not discussed widely despite how painful and challenging it can be to live with. Databases from the National Inpatient Sample (NIS) show that priapism is common, with approximately 5,000 admissions for this complication between 1998 and 2006 (Chrouser et al., 2011). Most of the 2,900 annual patients are adolescents. Although death rates associated with priapism are significantly low (approximately 0.71%), most patients have a low quality of life and face numerous challenges (Jamesetta, 2008), including chronic pain, acute painful episodes (also called painful events, painful crisis, or vaso-occlusive crisis), social stigma, depression, and anxiety. Even programs such as the National Health Service have been slow to respond to the complication (Demaso et al., 2004).

This research study strove to play a critical role in generating awareness among patients and in society more broadly regarding this SCD-related complication. In areas

where SCD is prevalent, this study may inform the development of effective psychological support services for patients. Given that the complication of priapism also affects a patient's family members, there is a clear need to provide them with quality psychological advice and assistance in coping with priapism (Panepinto, Pajewski, Foerster, Sabnis, & Hoffmann, 2009). By providing a deeper understanding of the lived experiences of individuals afflicted with priapism, this research study may challenge medical practitioners to improve the ways they handle patients with SCD, particularly those experiencing priapism (Addis, Spector, Shaw, Musumadi, & Dhanda, 2006).

Research has indicated that patients who suffer from SCD have often developed poor professional relationships with their doctors (Robinson, 1999). These deteriorating relationships between SCD patients and medical practitioners may be related to the subjective nature of pain and the fact that people react differently to conditions due to many factors, including the support of friends and family. Some health care providers doubt the veracity of complaints expressed by patients, which may compromise their concern for the patient's situation. The reason for providers doubting the veracity of complaints is often due to insufficient knowledge of sickle pathophysiology and triggers of events in SCD patients. Distrust between the patient and health care provider can lead to disparate and fragmented care (Robinson, 1999). The lived experiences described in this research study may therefore enable medical practitioners to better understand the issue of priapism in adolescents and encourage doctors to learn more effective approaches for identifying pain and counseling patients about such SCD-related symptoms.

This case study explored the lived experiences of 7 adolescent males suffering from SCD-related priapism. The research involved an interactive engagement with the patients to assess their situation and listen to each individual's story and perception of the complication (Burnett, Anele, & Derogatis, 2015). The researcher aimed to build an intentional rapport with the participants, acknowledge their struggles and challenges, and learn to relate with the patients in a nonthreatening manner.

Research Questions

The research questions for this qualitative phenomenological case study are:

1. What are the experiences of priapism among male adolescents with SCD?
2. How has priapism affected the lives of male adolescents with SCD?
3. What is the impact of the male adolescent's daily experience with priapism and SCD on the family?

Definition of Terms

Adolescents: Persons in the transitional phase of growth and development between childhood and adulthood. The World Health Organization refers to young people between 10 and 19 years old as adolescents (as cited in Erskine, 2011).

Lived experience: The subjective perception of a person's experience of health or illness (Nicholas, Picone, & Selkirk, 2011). Lived experiences differ by individuals according to the conditions with which they live (Adegbola, 2011).

Priapism: A persistent and painful erection of the penis (Chrouser et al., 2011). There are several different types of priapism. Ischemic priapism is a recurrent or "stuttering" priapism that occurs in males with SCD. Priapism occurs because of an inherited disorder

characterized by abnormally shaped red blood cells called sickle cell disease (SCD). After prolonged erection or unusually full bladder, some blood vessels in the penis are blocked by sickle cells, leading to intense pain (Erskine, 2011). Episodes of acute priapism are sometimes defined as an erection lasting longer than 3 hours. Fibrosis can develop in the penile shaft from longer episodes. Chronic blood-containing or fibrotic priapism develops from recurrent acute episodes. Acute-on-chronic priapism episodes can occur until fibrosis is complete and produces a permanently large heavy organ. Ejaculation can become premature and remains intact until lost by disuse. Fertility and testicular function remain intact.

Sickle cell disease (SCD): A group of hereditary disorders of red blood cells, characterized by abnormal hemoglobin, referred to as hemoglobin S or sickle hemoglobin (Adams-Graves et al., 2008). Hemoglobin, located in red blood cells, is the protein component responsible for transporting oxygen throughout the body. Sickle cell disease is an inherited disease; it is not contagious. People who suffer from SCD have inherited two abnormal hemoglobin genes, one from each parent (Powars et al., 2005). There are many forms of SCD and in all of them, at least one of the two abnormal genes usually causes the body to manufacture hemoglobin S. The most severe form of SCD occurs when the body forms two hemoglobin S factors. Other and more prevalent forms are hemoglobin SC and hemoglobin SBeta0 (Bennett, 2005).

Assumptions

This research study was based on the lived experiences of adolescent males with SCD and priapism. Although this research focused on 7 individuals, the researcher made several assumptions for the purposes of this study.

First, it was assumed that adolescent male patients with priapism behave in similar ways to those of the case study patients. Each person is different and responds to pain and disease in an individual way (Novy, 2011). The effects of the disease on an individual are based on many factors such as received moral support, family financial background, and the personality of the patient (Cita et al., 2016). It should be noted that some people exaggerate when they are in pain, some minimize, and some have difficulty thinking clearly; they thus present a challenge in determining the extent to which their narratives are truthful and accurate (Novy, 2011).

Second, it was assumed that because priapism is a complication of SCD that causes penile erection, it is associated with stigma that leads to feelings of guilt and shame (Panepinto et al., 2009). Such feelings can cause lower self-esteem and result in patients' avoidance of social engagements and problematic interpersonal relationships. Although it is widely accepted that illness is associated with low self-esteem, it is important to recognize individual differences in needs (Demaso et al., 2004). While reduced self-esteem is not an issue for all patients with SCD, severe instances can lead to deterioration in health and are a cause for concern (Bediako et al., 2016). The limitations of this study are considered in the following section.

Limitations of the Study

This qualitative phenomenological case study utilized semistructured interviews with adolescent males who have lived with priapism caused by SCD to learn more about their lived experiences. There were several limitations associated with this study.

First, the sample size of seven participants was too small to accurately capture the range of perceptions that other patients with SCD-related priapism might express (Schumacher, 2010). Moreover, given that adolescents range in age, and age can affect individuals' responses to many life issues (Pinckney & Stuart, 2004), adolescents may respond to SCD-related priapism in various ways depending on their age and maturity level. Therefore the results of this study are not generalizable to the broader population.

The information that the patients gave during the research study was self-reported, which is a limitation of this study because the information cannot be verified independently. Therefore, there is a possibility of bias and exaggeration (Sehlo & Kamfar, 2015). There is also the risk that the patient's information was not reported entirely accurately by the researcher. However, every attempt was made by the researcher to ensure accuracy. The significance of this study is discussed below.

Significance of the Study

One aim of this study was to increase awareness of SCD and priapism among the general population (Jenerette & Lauderdale, 2008). Given that so many people must live with SCD-related priapism, it is important to generate greater awareness of the complication and its challenging symptoms. This study reports facts about SCD and priapism with the hope of eradicating myths associated with them (Martin, 2011).

This study also aimed to help society better understand the unique struggles that adolescent males with SCD and priapism face (Bacsu & Metcalfe, 2012). Understanding the physical pain and desperation of an adolescent male with chronic pain caused by SCD and persistent, uncomfortable erections may encourage society to support other SCD male adolescents dealing with priapism (Dampier et al., 2010). Support from one's community can contribute to improved self-esteem and a faster rate of recuperation among this patient population (Tanabe, Dias, & Gorman, 2013).

The results of this study may be beneficial to both patients and doctors. Information provided in the study may prompt doctors to view priapism from the patient's perspective (Graff et al., 2012) and to devote more energy to developing supportive relationships with SCD and priapism patients and their families. Medical practitioners may also realize that they are in a position to offer appropriate counseling to this patient population (Khattab, Rawlings, & Ali, 2006). When patients feel that their community is aware of their condition and that their needs are recognized, their personal relationships and quality of life are likely to improve (Tanabe, Dias, & Gorman, 2013).

Chapter Summary

Research has shown that adolescent males with priapism often experience a poor quality of life. These individuals face many challenges, including low income levels, isolation, poor interpersonal relationships, frequent hospitalizations, and persistent pain (Shah & Hillinger, 2014). This study strove to build on extant research to continue to shed light on this complication and the experiences of adolescent males living with SCD-related priapism. In the following chapter, a review of literature is presented that considers the existing research in the field and places the current study within that

context. Chapter 3 presents the methodology and research design for the study. Chapter 4 presents the study findings, and Chapter 5 discusses the conclusions of the study.

CHAPTER 2

LITERATURE REVIEW

This qualitative phenomenological case study explored the lived experiences of adolescent male patients with SCD and priapism. Sickle cell disease is a common genetic blood disorder. The highest prevalence rates occur among Africans, Asians, and people of Mediterranean origin (Chrouser et al., 2011). This hemoglobin disorder, inherited from one's parents, is mainly characterized by vascular occlusion episodes, organ damage, and chronic hemolytic anemia, among other complications (Bacsu & Metcalfe, 2012). The United States has advanced in the management of the disorder through newborn procedures and prophylactic penicillin treatment of children with SCD (Bacsu & Metcalfe, 2012). However, in some African countries that lack modern medical technology advancements, the estimated numbers of children born with SCD that die in early childhood are alarmingly high (Mulumba & Wilson, 2015). Recent treatment regimens such as hydroxyurea have proved to be effective in the management of SCD in terms of prolonging and improving the quality of life. There is tremendous variation in the effects of SCD as a result of differences in genotypes, fetal hemoglobin levels, and steady state hemoglobin. Adolescents with SCD typically have multiple episodes of vaso-occlusive crises as a result of a rise in hemoglobin levels (Joice, Kates, Sopko, Hannan, & Bivalacqua, 2015). One of the major side effects of these episodes among adolescent males is priapism, the focus of this study.

This chapter provides an overview of priapism and its different manifestations, as well as complications associated with priapism and management protocols for priapism. Sickle cell disease is then discussed, including the types of SCD, complications

associated with SCD, management of SCD, and the psychological implications of the disease. A chapter summary concludes the chapter.

Priapism

Male adolescents with SCD experience nonsexual periods of prolonged penile erections, a complication known as priapism. These episodes are painful and may lead to impotence if left untreated (Joice et al., 2015). Priapism primarily occurs in the first few hours after sleeping. According to Joice et al. (2015), priapism affects nearly 10,000 males per year in the U.S., with a hospital admission rate of approximately 30%. Adolescents with SCD in particular often require medical assistance during this period of life. Compared to the average adolescent, patients with SCD experience delays in puberty averaging approximately 1-2 years. Those SCD patients with HbSS often experience even longer delays averaging approximately 2.5 years (Joice et al., 2015). In contrast, HbSC patients experience less delays due to higher hemoglobin levels but may experience more severe priapism as a result of those levels. Consequently, in addition to the physical symptoms of SCD and its complication of priapism, these patients are likely to be psychologically affected by the delayed growth and development in sexual characteristics (Sehlo & Kamfar, 2015).

According to pediatric urologists, priapism is among the most common and difficult clinical complications associated with SCD (Wang et al., 2016). Statistics indicate that SCD accounts for about 65% of all cases of priapism in children (Wang et al., 2016). Approximately 89% of all men with SCD reported at least a single episode of priapism before the age of 20 (Wang et al., 2016). Medical records indicate that between 2004 and 2012 there was a substantial decline in the number of priapism patients

admitted in hospitals. This can be attributed to improved outpatient management strategies resulting in fewer adolescents with SCD likely to develop priapism (Wang et al., 2016). Improved preventative medication regimens have also contributed to lower rates of hospital admissions (Wang et al., 2016).

It is common for adolescents with SCD-related priapism to experience stigmatization and isolation, especially during recreational activities. This social alienation often leads to anxiety that results in co-occurring psychological conditions such as depression, academic underachievement, and rebellious behavior (Sehlo & Kamfar, 2015). According to Sehlo and Kamfar (2015), SCD complications including priapism can involve a host of cognitive, psychological, and psychosocial comorbidities. As a chronic illness, SCD can result in other depressive disorders, especially during the teenage years. Although not all studies have shown a significant risk of childhood depression in SCD cases, recent studies have shown that more than 50% of adolescents diagnosed with SCD were also diagnosed with either dysthymia or major depression (Sehlo & Kamfar, 2015). Dysthymia accounts for nearly 90% of childhood depression cases among adolescents with SCD (Sehlo & Kamfar, 2015).

Sickle cell disease and its associated complications also impact the quality of life of patients. Adolescents with SCD generally have a lower health-related quality of life (HRQL) compared to their peers without the disease (Sehlo & Kamfar, 2015). However, social support systems have been shown to be impactful, as SCD children and adolescents with a strong social support system generally have few adjustment problems. Moreover, SCD patients involved in self-help support groups tend to report fewer psychological symptoms. Sehlo and Kamfar (2015) thus proposed that adolescents with

SCD be encouraged to increase their participation in such support groups as a way of reducing feelings of depression. There are several different types of priapism, which are discussed in the following section.

Types of Priapism

Priapism refers to the process during which abnormal blood flow to the spongy tissues in the penis can cause a nonsexual erection lasting up to four hours (Olujohungbe & Burnett, 2013). Priapism is assigned to the hemolytic-endothelial dysfunctions phenotype, and flow of blood in both macro- and micro-circulation is caused by blood rheology (Olujohungbe & Burnett, 2013). The influences of blood circulation include aggregation properties, red blood cell deformability, and blood viscosity (Cita et al., 2016). The pathophysiology of SCD-related priapism remains unclear (Cita et al., 2016). The complication results from sludging of the sickle red blood cells in the cavernous sinuses (Olujohungbe & Burnett, 2013), which is assumed to interfere with venous outflow (Cita et al., 2016). Further sickling of red blood cells may result from acute cavernosal hypoxia, causing cavernosal smooth muscle paralysis and a priapic event (Olujohungbe & Burnett, 2013). However, recent studies have suggested that changes in the physiology of erection signaling pathways, such as the PDE5 pathway, as well as impairment of the adenosine signaling, can lead to priapism episodes (Cita et al., 2016). The two main types of priapism are: (a) *low-flow priapism*, also known as *ischemic priapism*, and (b) *high-flow priapism*, also known as *non-ischemic priapism*. A third type of priapism, known as *stuttering or intermittent priapism*, has also been reported (Olujohungbe & Burnett, 2013).

Low flow (ischemic) priapism. Ischemic priapism does not have a defined cause and is found primarily in adolescents and men with SCD, malaria, or leukemia (Cita et al., 2016). The obstinate, nonsexual erection is characterized by abnormal cavernous blood gasses. In some cases, there is little or no cavernous blood flow (Cita et al., 2016). The cavernous blood gasses can be hypoxic, hypercarbic, or acidotic. This type of priapism is also characterized by a corpora cavernosa that is rigid and tender to palpation (Cita et al., 2016). The complication is painful, and a host of etiological factors can lead to failure of detumescence mechanisms. Therefore, it is an emergency complication that requires medical attention. Although ischemic priapism is self-resolving, whereby the penis returns to a flaccid and nonpainful state, conditions such as persistent penile edema, partial erections, and ecchymosis may occur and mimic priapism (Cita et al., 2016). This type of priapism can be resolved by color duplex ultrasonography, which is useful in measuring blood flow and cavernous blood gasses. Despite its prevalence, ischemic priapism lacks an optimum therapeutic intervention (Cita et al., 2016).

High flow (non-ischemic) priapism. Non-ischemic priapism is a nonsexual erection that results from unregulated flow of blood in the cavernous arteries. However, unlike low-flow priapism, cavernous blood gasses in non-ischemic priapism are neither hypoxic or acidotic (Cita et al., 2016). This type of priapism also differs from the low-flow type in that the penis is neither fully rigid nor painful. The most described etiology in non-ischemic priapism is antecedent trauma. As opposed to low-flow priapism, this type of priapism is not an emergency complication requiring medical attention (Quinn, Rogers, McCavit, & Buchanan, 2010). Resolution of this type of priapism is a return of the penis to a completely flaccid state.

Stuttering (intermittent) priapism. This type of priapism refers to painful nonsexual erections that occur repeatedly, with intervening periods of detumescence (Cita et al., 2016). The main treatment option for this type of priapism involves care to prevent future episodes. Cita et al. (2016) argued that this form of priapism, which occurs in brief clusters, may cause inflammatory reactions as a result of penile tissue ischemia, possibly resulting in erectile dysfunction, penile fibrosis, and impotence.

According to Ogwumba et al. (2015), in more than half of the cases of SCD patients who previously suffered from stuttering priapism, low-flow priapism presents itself subsequently. In a bid to determine the etiology, presentation, management, and outcomes of ischemic priapism, Ogwumba et al. (2015) studied 27 patients who were undergoing ischemic priapism management in three hospitals in South East Nigeria between January 2000 and December 2010. Clinical data such as clinical features, interval and onset of presentation, and type of priapism were retrieved. Of the 27 patients, only 15 had a complete clinical record. The mean age of the patients was 30.5 years; 9 were students. Eight of the 15 patients had SCD and had previously suffered from stuttering priapism before advancing to ischemic priapism. Four had unidentified causes (Ogwumba et al., 2015). Six had taken herbal medications before the presentation of medical attention. After the initial resuscitation consisting of intravenous fluid and aspiration, detumescence was immediate in all but one patient, in whom detumescence was delayed. Recurrence of tumescence occurred in three patients, marked by expression of blood in the shunt sites (Ogwumba et al., 2015).

Based on the outcomes of the aforementioned study, it is evident that low-flow priapism is the most common type of priapism among SCD patients, with previous

stuttering priapism accounting for approximately 50% of all cases (Ogwumba et al., 2015). Timely treatment of the complication significantly reduces the possibility of severe erectile dysfunction. Ogwumba et al. (2015) suggested use of the Al-Ghorab shunt, which provides ischemic priapism patients with immediate relief. Given the severity of ischemic priapism among SCD patients, public awareness is critical to facilitate early intervention and minimize the probability of severe erectile dysfunction (Ogwumba et al., 2015). Ischemic priapism prevention measures are thus vital for patients with SCD. The various complications associated with priapism are explored below.

Complications of Priapism

Sickle cell disease is characterized by episodic acute complications during adolescence that may continue to adulthood (Dampier et al., 2010). Adolescents with the disorder display symptoms of chronic multi-organ failure and present with various complications. Major complications associated with SCD in adolescents include pain as a result of priapism and sickle erythrocyte vaso-occlusion (Dampier et al., 2010). Most patients with priapism are likely to be found in a tertiary referral center, since they often procrastinate seeking medical attention until after complications have become severe due to stigma associated with priapism (Furtado et al., 2012).

Complications associated with priapism range from clinical and medical complications to psychological complications (Erskine, 2011). Physical complications include penile gangrene, severe erectile dysfunction, and penile deformity (Burnett et al., 2015). Other chronic complications that are associated with SCD in adolescents include slight dilation of the left ventricle, respiratory function difficulties, increased flow

velocity of cerebral arteries (due to damaged vessels), cholelithiasis, and recurrent priapism (Silva et al., 2014). Patients with SCD are prone to cold and tend to be susceptible to other health challenges including infection (Asakitikpi, 2015). Adolescents with SCD and priapism also experience psychological complications. These patients are particularly prone to anxiety, primarily due to the perception that they are judged negatively in their communities due to what many perceive as an embarrassing condition (Robinson, 1999). According to Asakitikpi (2015), meanings assigned to social behavior mediate people's actions toward an external stimulus. Therefore, the perception of being judged, in this case due to their nonsexual penile erections, usually causes adolescents with priapism to become more reserved and to fail to interact with peers, and even withdraw from social spaces (Asakitikpi, 2015). The physical complications of penile gangrene and erectile dysfunction are addressed in what follows.

Penile gangrene. Penile gangrene is a rare complication of priapism. After the Al-Ghorab shunt has been carried out and subsequent complete detumescence, ischemic changes may occur on the distal part of the penis about four days post intervention (Ogwumba et al., 2015). Penile gangrene has been attributed to a wide range of factors, including self-inflicted penile strangulation, chronic renal failure, dialysis, and diabetes mellitus. Most cases of complicated priapism are usually idiopathic in origin and generally of the ischemic type. Treatment of penile gangrene involves active resuscitation of the patient and urgent decompression of the intracavernosal pressure for complete detumescence.

Erectile dysfunction. Erectile dysfunction occurs as a result of narrowing penile blood vessels (Anele & Burnett, 2015). Erectile dysfunction is a common condition

among men, particularly those between 40 and 70 years old. However, it can also occur among patients with priapism post intervention; in most cases, the condition is a long-term risk after intervention (Anele & Burnett, 2015). The warning is usually indicated on the discharge instruction sheet and documented in the chart. Low-flow priapism has a higher risk of erectile dysfunction compared to high-flow arterial priapism. Management options for priapism are considered in the next section.

Management of Priapism

Broadly speaking, the traditional method and aim of managing priapism has been centered on achieving a sound clinical resolution to return an erectile penis to its normal flaccid state (Burnett et al., 2015). Surgical procedures used for treating priapism include intracavernosal sympathomimetics, aspiration, and shunt procedures. Pharmacologic approaches are also extensively used to treat ischemic priapism. However, according to Burnett et al. (2015), treatment options that are based solely on resolving abnormal erections usually fail to estimate the scope and magnitude of the health problem. Given the number and types of complications associated with SCD-related priapism, it is important to develop more comprehensive treatment options that can balance between monitoring the holistic well-being of the SCD patient and continuously assessing the success of clinical treatment.

Standard management of priapism also involves efforts to reduce episodes through fluid management and bladder emptying. Given that an overfull bladder precipitates or prolongs erections, and bladders are prone to overfill at night, management of priapism includes fluid restriction after supper and voiding the bladder at bedtime. If the patient is susceptible to acute episodes of priapism, they are trained to set their alarm

clock to wake up two sleep cycles after bedtime (approximately 3 hours), in addition to voluntary emptying of the bladder during the night. This protocol also commonly includes bedtime sympathomimetics.

Adolescent males with SCD require constant evaluation, medical care, and treatment, especially in priapism cases. These requirements can exert an adverse impact on family income and lead to poor quality of life for the adolescent (Panepinto et al., 2009). Panepinto et al. (2009) conducted a study to determine the effects of SCD on adolescents and its subsequent impact on quality of life and family income involving children with SCD and a control group of children without the disorder. Participants completed a self-report questionnaire or a PedsQL generic core scale during a clinical visit. The results suggested that, taking into consideration all variables including age of the patient, family income, and comorbidities, the children with SCD had greater odds of poor HRQL (Panepinto et al., 2009).

By evaluating HRQL, it is possible to determine the best assessment of disease outcomes on patients in terms of disease complication, the course of the illness, and treatment procedure. Generally, HRQL among adolescent males with SCD is poor. However, sufficient details regarding the effects of the disease itself on the level of HRQL do not exist because of co-occurring factors, such as level of family income.

Photovoice, a method for patients to share their experiences with a disease through photo stories, may be an effective management option for SCD-related priapism as well. Valenzuela et al. (2013) conducted a photovoice pilot among young people with SCD. The researchers suggested that photovoice is significant as a participatory method for adolescents with priapism to share their perspectives. This practice of sharing their

experiences appears to help patients cope with the difficulties of SCD and its related complications, including the recurrent, chronic, and sporadic pain resulting from priapism episodes (Valenzuela et al., 2013). Some of the experiences of adolescents with SCD included a tendency to miss school, especially during the commencement of treatment. In addition, individuals undergo ongoing treatment procedures such as monthly transfusions, medical procedures like MRIs, and other routine screenings including vision and hearing tests, all detracting from school time (Valenzuela et al., 2013). The changes in lifestyle required to manage the disorder, such as taking medications regularly, avoiding extreme temperature changes, drinking fluids, and eating a balanced diet, also prove difficult for adolescent patients. Compliance with these requirements leads to lower HRQL, especially among adolescents who cannot interact normally with peers. Valenzuela et al. (2013) found that photovoice helped to promote engagement and understanding of the condition among adolescents with SCD, which may contribute to increased self-esteem.

Another management approach suggested to minimize the impact of fragmented care on patients with chronic disease like SCD is developing positive relationships between health care providers and patients (Adegbola, 2011). Given the challenges associated with pain management among adolescents and adults with SCD, especially during episodes of priapism, improved communication and interactions with the treating doctors would likely be beneficial to patients' well-being (Adegbola, 2011). According to various studies, patients with chronic pain tend to be treated differently and nontherapeutically by their health care providers (Adegbola, 2011; Thomas, 2006). In Thomas's (2006) study, health care professionals described patients with SCD as

generally difficult and communication with them as challenging. Health care providers' communication skills and cultural awareness training can have significant outcomes, especially on their confidence to communicate with SCD patients (Thomas, 2006). This, in turn, can improve the quality of care and general management of the disease and its complications.

The most applicable approach to treatment of chronic pain resulting from SCD is holistic evaluation of the pain based on the patient's perspective (Adegbola, 2011). The traditional pain grading system is designed for evaluation of acute pain and may fail to reflect the patient's intended response. Therefore, open-ended questions as part of a qualitative inquiry to assess the level of pain are particularly preferred for use with adolescents with priapism (Adegbola, 2011).

Extended family and a strong social support system has also been found to aid in the management of the psychological complications associated with SCD and priapism among adolescents. At this stage of development, responsibility for one's health starts to shift from the parent to the adolescent, which can have adverse effects on HRQL. According to Telfair and Gardner (1999), adolescents with priapism who perceive that they have a supportive environment in the management of the complication tend to experience more positive biopsychosocial outcomes, evaluated through reported cases of physical and psychological functioning. Graff et al. (2012) examined the impact of family communication on the general welfare of adolescents with SCD and priapism, concluding that extended family can provide a critical network and resources for such adolescents.

Other research has corroborated Graff et al.'s (2012) findings regarding HRQL among adolescents with SCD and associated complications. A study evaluating the lived

experiences of adolescents with SCD in Kingston, Jamaica applied a descriptive qualitative design through semistructured interviews with six participants (Forrester, Barton-Gooden, Pitter, & Lindo, 2015). Study results suggested that adolescents who were more actively involved in social activities and parties, and who had strong family, school, and peer support, reported significantly higher levels of HRQL.

Various other coping strategies, such as surfing the Internet, praying, and watching television have been shown to be effective for managing the psychological complications of priapism in adolescents (Forrester et al., 2015). In addition, family members' knowledge of the complication is an important determinant of communication. Compared to communication between adolescents with SCD and their siblings, communication between adolescents with SCD and their parents was more effective because parents tended to have a deeper knowledge of the disease. The parents' response to the disease and their degree of communication about SCD with an adolescent child is vital to the diagnosis of the condition and the patients' overall experience (Graff et al., 2012). Harrison et al. (2016) also reported that persons who learn about SCD from their families have a higher level of knowledge about the condition, which is an important factor in the management of priapism in adolescents with SCD. An overview of research on SCD and its associated complications and management options is presented in the following section.

Sickle Cell Disease

Children and adolescents with SCD in the United States are primarily African Americans (Panepinto et al., 2009). The disorder affects one African-American in every 500 live births in the United States (Powars et al., 2005). Research has shown that

poverty is a major risk factor in determining HRQL in patients with the disorder. Sickle cell disease does not have a cure and medical care for adolescents with the disorder is palliative, focusing on pain management (Telfair & Gardner, 1999). The unpredictable complications of SCD negatively impact afflicted adolescents biologically, psychologically, and in social development (Telfair & Gardner, 1999).

Despite the prevalence of the disease among African Americans, awareness of SCD varies broadly across the population. Harrison, Walcott, and Warner (2016) found that knowledge of the disorder among African-American women was critical to the management of SCD and priapism among adolescent patients. However, the level of misinformation about the condition remains high (Harrison et al., 2016). This section presents the different types of SCD, complications associated with the disorder, management options for SCD, and the psychological implications of the disease.

Types of SCD

The four types of SCD result from gene mutation of the hemoglobin in red blood cells. Hemoglobin SS disease (sickle cell anemia) is the most common and severe type of SCD, resulting from inheritance of the hemoglobin S gene from both parents (Bennett, 2005). The second most common type, less severe than hemoglobin SS disease, is hemoglobin SC disease, resulting from inheritance of the Hb C gene and Hb S gene (Bennett, 2005). The third type of SCD is Hb SBeta 0 Thalassemia, which is similar to SS disease because there is no HbA. The fourth type is SBeta+ Thalassemia, which is relatively mild because there is 20% HbA. Common complications associated with SCD are explored below.

Complications of SCD

Apart from priapism, other complications can occur as a result of SCD, including eye problems as a result of the blockage of vessels that supply the eyes. Severe forms of anemia can occur as a result of hemolyzing sickle cells. A hand-foot syndrome may occur wherein both hands and feet swell due to vessel blockages by sickle-shaped red blood cells (Bennett, 2005). Other complications include delayed growth, whereby sexual maturation takes longer than normal. The breakdown of red blood cells and high levels of bilirubin can also cause gallstones. The disease and its related complications are managed in various ways.

Management of SCD

In SCD management, health professionals are in a critical position to reduce the severity of the disease and other complications, such as priapism. It is vital for health care providers to understand the manifestations of the disease, life-threatening complications, and optimal management strategies. Jenerette and Lauderdale (2008) outlined the lifelong patterns of SCD, which can involve pain episodes, anemia crises, infections, and stroke. Patients with SCD can live normally if they are accorded the right kind of care and treatment (Jenerette & Lauderdale, 2008). In therapeutic strategy, prophylaxis is of paramount importance in averting adverse outcomes of the disorder and improving the level of HRQL (Khattab et al., 2006).

Adolescents with SCD-related priapism should undergo all defined prophylactic strategies to manage the complication, including lifestyle intervention. For instance, patients should avoid high altitudes, cold, alcohol, and smoking (Khattab et al., 2006). Other major contributing factors to high levels of morbidity and mortality in adolescents

with SCD are viral and bacterial infections. Therefore, health care providers can improve the quality of life of adolescents with SCD-related priapism by administering vaccinations against meningococcal, pneumococcal, and Hemophilus bacterial infections (Khattab et al., 2006). Administration of oral penicillin is an effective method for reducing infection rates (Khattab et al., 2006). Another important prophylactic strategy for adolescents with SCD-related priapism is administration of folic acid in the presence of severe chronic hemolysis (Khattab et al., 2006). Hydroxyurea is administered to increase levels of fetal hemoglobin (HbF). Many acute episodes and hospital admissions can be reduced significantly with this prophylactic strategy (Khattab et al., 2006). However, hydroxyurea should not be administered during pregnancy.

Apart from hospital care, community care plays an important role in management of SCD-related complications such as priapism among adolescents. Community care usually takes place at home with input by pediatricians, hemoglobinopathy counselors, and community nurses. The primary role of these health professionals is to educate the community and the relatives of adolescents with SCD-related complications regarding ways to manage the disorder. Apart from direct medical care, community care addresses chronic medical and social problems resulting from SCD-related complications (Khattab et al., 2006), including challenges surrounding upholding employment and education-related obligations. A combination of all treatment and management strategies significantly improves patients' level of HRQL.

In the course of diagnosis, treatment, and management of the disorder, health care providers' interventions should be primarily concerned with coping strategies. Common coping strategies among SCD patients include prayer and participation in social activities

(Webb, 2010). Successful management of this lifelong disorder depends on patients' levels of independence and the support provided to them, especially during adolescence.

Factors determining the pediatric to adult care transition among SCD adolescents are poorly understood (Newland, 2008). The primary difficulty facing adolescents with SCD is inadequate skills for managing their lives as they undergo this transition in their medical care simultaneous to their personal transition into adulthood (Newland, 2008), which involves both physical and psychological change (Powars et al., 2005). In particular, new relationships and behaviors cause changes in both internal and external experiences. One method of managing adolescents with SCD and priapism is to provide transitional programs to ensure that they are well-positioned to acquire developmentally appropriate medical care (Newland, 2008). In the transition program proposed by Newland (2008), decision-making is transferred first from the parent to the adolescent–parent unit. Later, decision-making capacity transfers fully to the adolescent (Newland, 2008).

With recent medical advances and new management methods for the disorder, the life expectancy for individuals with SCD has significantly improved, encouraging research on the underlying psychosocial factors that influence the quality of life among adolescents with the condition (Pinckney & Stuart, 2004). Interpersonal factors are among the most important psychosocial factors for adolescents with SCD, including difficulties in assertive communication, decreased coping strategies, and challenges of processing stress (Pinckney & Stuart, 2004). Other psychosocial problems affecting adolescents with SCD include family factors such as family support, parent–child relationship problems, cohesion, organization, and control (Pinckney & Stuart, 2004).

Demographic variables that contribute to poor quality of life among adolescents with SCD include age, gender, and socioeconomic status. All of these factors adversely affect adjustment, and disease morbidity is significantly related to such adjustment difficulties among adolescents (Pinckney & Stuart, 2004).

In a similar study evaluating the nature of pain among adolescents with SCD-related priapism, Booker, Blethyn, Wright, and Greenfield (2006) conducted a thematic analysis of material from focus groups to understand barriers described by participants. Patients reported that feelings of isolation represented the most critical factor affecting pain management in SCD. For them, the “crisis” pain of SCD resulted from failure to be heard, as well as limited social support (Booker et al., 2006). The study also found that certain factors bar adolescents from seeking early treatment in SCD-related priapism, including anxiety and fear of being judged, as well as a lack of understanding of SCD. The ongoing psychological implications of SCD are considered in further detail in the following section.

Psychological Implications of SCD

Most patients with SCD living beyond early childhood report psychological and social factors that make adjustment particularly difficult (Bediako et al., 2016).

Psychological and mental health complications occurring as a result of SCD-related priapism include feelings of embarrassment, despair, anxiety, and isolation (Burnett et al., 2015).

Social stigma, the phenomenon whereby an individual with a socially unfavorable attribute is rejected by society, has become a central concern for patients with chronic illnesses including SCD. Yet, there remains limited research on the influence of social

stigma on the health outcomes of adolescents with SCD. In particular, external negative self-evaluation by SCD patients causes feelings of vulnerability, which result in an increased disease burden due to delays in seeking medical and psychosocial support (Bediako et al., 2016). Among adolescents with ischemic priapism, complications may arise from premature treatment termination as a result of stigma and related issues. In a study to evaluate the effects of stigmatization on SCD patients, Bediako et al. (2016) administered the Measure of Sickle Cell Stigma, a recently developed standard tool, to 262 patients. The study produced a consistent outcome, suggesting that a greater level of social stigma led to more frequent association with medical care for SCD patients. Results also showed a significant association between perceived stigma, pain-related health utilization, and the perceived disease (Bediako et al., 2016).

Certain prominent physical complications of SCD, such as leg ulcers and jaundiced eyes, may be sources of stigma, resulting in feelings of anxiety and shame, especially among adolescents (Bediako et al., 2016). Such feelings can significantly hinder interpersonal and other social interactions. Although SCD can be treated at early presentation with therapeutic intervention, awkwardness and embarrassment associated with the condition prevent many adolescents from reporting it. The racial association of SCD may also lead to stigma given that most people perceive the condition as one experienced primarily by persons of African descent. Bediako et al. (2016) suggested that SCD stigma-related factors such as disclosure concerns, internalized stigma, and expected discrimination play a major role in preventing adolescents with priapism from receiving medical attention.

Beyond stigma, numerous other factors can influence the quality of life of adolescents with SCD. According to Graves, Hodge, and Jacob (2016), such factors may involve the physical environment surrounding these adolescents, including their neighborhoods, homes, and schools. Other factors that adversely affect their levels of HRQL include socioeconomic distress, unemployment, and school dropout (Graves et al., 2016). Graves et al. (2016) concluded that, as clinicians focus on managing painful episodic exacerbations in SCD, especially in the case of priapism, it is vital to pay attention to school-related factors and to offer psychosocial support.

Chapter Summary

Sickle cell disease-related priapism among adolescents has significantly contributed to a reduced HRQL for these individuals. Priapism and other complications of SCD result in both the deterioration of the physical body and in psychological challenges. According to Booker et al. (2006), adolescents with priapism generally experience feelings of isolation and demonstrate poor coping strategies such as anger, active avoidance of service use, and aggression. The researchers suggested forming pain discussion self-help groups and providing specialized treatment and other services (Booker et al., 2006).

The strategies used by adolescents with SCD-related priapism in coping with pain, moreover, have a direct correlation with the level of support received from family and relatives. Positive coping strategies adopted by adolescents with priapism include problem-solving, seeking information, social support, and positive self-statements (Graves & Jacob, 2014). Given the notable psychological implications of SCD-related priapism in adolescents, additional research is needed to shed further light on the

complication and gain a better understanding of patients' lived experiences with SCD and priapism. The following chapter discusses the research design and methodology utilized in this study.

CHAPTER 3

RESEARCH METHODOLOGY

Phenomenology

This chapter describes the methods used in this qualitative phenomenological case study on priapism among adolescent males with SCD and priapism. Working closely with adolescent males to study their experience with priapism may offer new insights that can be implemented in order to improve this population's health conditions. Studies have shown that adolescent males with SCD who are suffering from priapism are likely to experience psychosocial dysfunction and poor academic performance as well (Ohene-Frempong et al., 1998). Building on previous phenomenological studies of priapism in male adolescents with SCD (Chopra, Harley, Lahiff, & Eskenazi, 2014), this study was designed to collect phenomenological descriptions from adolescent males with SCD and priapism. The study utilized measures that have not been employed in past research to explore the psychosocial functioning of American adolescent males with SCD and priapism in southern California. This chapter addresses the research design, study sample and participants, data collection and analysis, validity and reliability, ethical considerations, and the role of the researcher.

Research Design

This qualitative study was exploratory since the body of existing research on priapism among adolescent males with SCD remains small. This research employed a case study design to gain information about priapism in adolescent males with SCD living in southern California. A case study design using semistructured interviews was

employed because it was expected to increase the internal authenticity of the data in comparison to other techniques (Adeyoku et al., 2002).

The research design relates to the decisions of the researcher as they evolve from a general idea to specified data collection and analysis methods (Creswell, 2009). The researcher referred back to this idea throughout the research process to ensure that the data addressed the research questions as unequivocally as possible (Creswell, 2009). A consistent research design minimizes the probability of drawing incorrect causal inferences from data (Creswell, 2009).

In this qualitative research study, convenience sampling was used to identify SCD male adolescents with priapism in the region of southern California. A level of selection bias associated with convenience sampling is unavoidable given that participants respond voluntarily to advertisements for the study (Polit & Beck, 2012).

The qualitative methodology of this study involved a phenomenological approach whereby the researcher identifies a specific source of information, in this case SCD adolescents and support groups in southern California (Creswell, 2009). With this approach, the selected case study participants related historical information regarding the origin of a problem which, in turn, helped the researcher to understand this population's life experiences (Creswell, 2009). The phenomenological approach was chosen in order to explore how priapism affects adolescent males with SCD by developing knowledge through the description of lived experiences, with the participants describing how priapism affected their lives both psychosocially and academically. Other qualitative strategies might be less effective in providing a detailed and comprehensive understanding of how priapism affects adolescent males with SCD. For instance,

grounded theory provides broad information by generating a theory about the life experiences of the participants but does not provide specific information (Creswell, 2009). In other words, grounded theory cannot provide adequate information about what adolescent males with SCD are experiencing. A case study design was deemed appropriate for this study because it allowed an in-depth exploration of the lived experiences of a small sample of adolescent males with priapism, thus enabling deeper data collection.

Advantages of using a qualitative phenomenological approach include the ability to collect historical information from participants, with the researcher having control over the questions posed to participants and the absence of the need to observe participants directly (Creswell, 2009). Limitations to this approach include the provision of extraneous, indirect, or incomplete information, which may result in researcher bias.

Sampling Method

In qualitative research, the research objective and characteristics determine the number of participants to recruit for the study. This research study used a convenience sampling method (Creswell, 2009; McKenzie, Neiger, & Thackeray, 2008) to select the sample. Convenience sampling gives all members of the study population a chance of selection to participate in a study. Furthermore, convenience sampling reduces the chance of irrelevant participants in study samples (McKenzie et al., 2008). This sampling strategy was appropriate for the present study since it effectively locates samples in hidden populations not easily accessible through other sampling strategies (Creswell, 2009; Marshall, 1996). The sample frame consisted of seven adolescent males with SCD

and priapism between the ages of 16 to 19 years old who were participants in priapism and SCD support groups.

Sample Criteria

The inclusion criteria for this study were:

- Adolescent males between the ages of 16 to 19 years old
- Diagnosed with SCD and priapism
- Ability to effectively narrate their experiences with SCD and priapism
- Demonstrated willingness to participate in this study

Setting

The choice of setting for the interview is very important as it can have a positive or negative effect on the data collection process (Creswell, 2009). The researcher and participant must come to a mutual agreement on a setting suitable for both. The participant must be at ease and comfortable in order to give a productive interview. Ideally a quiet location is the best choice for conducting an interview, such as the group study room of a library or a private office setting. An outdoor location such as a park or restaurant can be chosen but distractions such as loud music or people playing and moving around can affect the participant's chain of thoughts. Depending on each participant's preference and comfort level, the researcher met with potential participants either in a public library, physician's office, or local restaurant or coffee shops to complete the demographic questionnaire and semistructured interview.

Data Collection

This section describes the procedures for collecting data. Every participant completed a self-administered questionnaire (Appendix A) to provide demographic information before the interview, including age, ethnicity, grade and level of study in school, hemoglobinopathy (SCD type), and patient's history of health (Kato et al., 2006). The researcher also recorded his own observations throughout the course of the interviews.

The interviews required consent by participants (Appendix B). Qualitative data was drawn from semistructured face-to-face interviews guided by an interview protocol (Appendix C) designed to explore the experiences of adolescents with SCD-related priapism. The protocol incorporated open-ended questions, allowing for elaboration by the interviewee (Creswell, 2009). The researcher prepared the interview questions, which were reviewed and approved by his committee members.

Interview questions were categorized into three dimensions, which were uniform for all participants: (a) the participant's general information, (b) psychosocial relations of the participant, and (c) the general life and health of the participant with respect to SCD (Mantadakis, Cavender, Rogers, Ewalt, & Buchanan, 1999). However, the depth of probing depended on how each participant answered the questions, since some gave responses that required additional follow-up questions for the sake of clarity. Interviews were audio recorded.

During the interview, the researcher also took notes on the participants' nonverbal cues and activities using a prepared template. This information included date, time, setting, and observations regarding the participant's gestures and facial expressions, as

well as insights from the researcher. Two hours were allotted for collection of data from each participant. Completion of the questionnaire took around 6 minutes, while the oral interviews took between 45 to 60 minutes.

Consideration of ethical issues in research is critical (Lorig & Holman, 2003; Motacki & Burke, 2011). In this case, the researcher observed university guidelines to ensure ethical conduct of the study. The researcher presented a consent form for signature (Appendix B) to all potential participants. The form stated that participants were not under duress to participate and that they consented to do so by their own free will (Huang, O'Connor, Ke, & Lee, 2016). Before the participant signed the consent form, the researcher explained the nature and purpose of the questions and overall data collection (Melgoza, Mennel, & Gyeszly, 2002). Confidentiality of all data collected was ensured to encourage smooth exchange of information without feelings of coercion (Melgoza et al., 2002).

When research participants are ensured of confidentiality and anonymization of study data, they tend to be truthful in providing information (Petrova, Dewing, & Camilleri, 2016). However, when they are uncertain of the state of confidentiality and anonymity, they tend to withhold information that may be useful to the research study. In consideration of confidentiality and trust, participants did not provide identifying information except gender (Petrova et al., 2016), since such information might be seen as a breach of privacy and might lead to the participants withholding information. Each participant was assigned an ID number for the purpose of collecting and analyzing data. Data is already stored under lock and key to prevent unauthorized use by anyone other than the researcher; this protocol is explained on the consent form.

Data Analysis

This study was designed to address three research questions:

1. What are the experiences of priapism among male adolescents with SCD?
2. How has priapism affected the lives of male adolescents with SCD?
3. What is the impact of the male adolescent's daily experience with priapism and SCD on the family?

Data analysis in this study was conducted in four steps and did not require use of software programs. Analysis of data was based solely on note taking and audio transcribing of interviews. The type of data collected in this study required pooling themes, patterns, categories, and relationships, all of which could be identified by the researcher without the assistance of software programs (Polit & Beck, 2012).

The first step in data analysis is to transcribe the recorded interview data. Verification of the transcript is necessary to remove the possibility of bias (Amankwaa, 2016). A copy of the transcribed interview was issued to the respective interviewee to verify accuracy. After transcript verification was completed for all participants, information was grouped based on themes, patterns, and categories to organize the responses for similarities and differences (Amankwaa, 2016).

The second step is to analyze the detailed notes of the transcripts with specifications on the document identifying the participant. Each participant received an ID number (Amankwaa, 2016). Data was organized based on the ID number, question number, responses, and code. The question number identifies the given question and the code describes the type of response. Responses were organized in tabular form (Amankwaa, 2016). Three identifying codes were used: depression level, anxiety, and

oppositional defiant behavior. The first determines whether depression levels are high among adolescent males with priapism and SCD. The second determines whether levels of anxiety are high among adolescent males with priapism and SCD. The third determines levels of oppositional defiance behavior among adolescent males with priapism and SCD.

The third step in data analysis is to review participant responses to identify themes, patterns, and relationships (Islam & Tanasiuk, 2013). Themes that are recurrent in participant responses are identified; patterns are recognized by the manner in which participants give responses, and relationships identify how responses relate to one another.

The fourth and final step involves summarizing the data, including the themes, patterns, and relationships that were identified (Islam & Tanasiuk, 2013). The principal focus is the identified themes which reflect the central, recurrent ideas that emerged from the participant responses.

Participants

The following procedures were employed to recruit study participants. Participants were recruited through a single SCD support group located in Loma Linda, CA. The target population for this support group is families with SCD. It is comprised mainly of African-American families. The purpose of this group is to provide financial assistance, emotional support, peer mentoring and visitation, a discount prescription program, a discount dental program, grant and scholarship programs, and health and fitness activities to families and individuals diagnosed with SCD.

The researcher initially approached the SCD support group coordinator via telephone to seek permission to recruit male adolescent SCD patients who have experienced priapism between the ages of 16 and 19. A formal letter describing the intent and design of the study (Appendix D) was subsequently sent to the support group coordinator, requesting assistance in recruiting participants by distributing flyers (Appendix E) to support group members. After receiving permission from the program coordinator, potential participants were met in public libraries, local restaurants, coffee shops, delis, or a physician's office to complete demographic questionnaires and semistructured interviews.

Flyers were posted in various locations with contact information for potential participants. The flyers described procedures and offered compensation for participating in the study. Prospective participants made contact with the researcher via the telephone number or email address on the flyer for participant screening and to schedule interviews with eligible participants.

Validity and Reliability

This study required specific methods for establishing reliability. In qualitative research, reliability is measured by the degree of credibility, transferability, dependability, and conformability of the research (Ngulube, 2015).

Credibility refers to the extent of validity of a study (Ngulube, 2015). Credibility in this study was ensured by conducting a peer review of the findings, which was completed by the researcher's committee members. The codes identified in data analysis were monitored by the primary researcher to ensure that there was no drift in the meaning of codes or use of codes for other meanings. Validity was supported by restricting contact

between participants and ensuring that participants were not privy to information provided by other participants.

Validity can be threatened by social desirability, which may occur when participants give certain responses to please the researcher (Ngulube, 2015). To avoid such responses, participants were assured that there were no pressures or expectations for answering the interview questions (Ngulube, 2015).

Transferability measures the degree to which research findings can be applied to other contexts. In this case, transferability was ensured by verifying that participants have received a diagnosis of priapism with SCD (Cita et al., 2016).

This collection of measures to test trustworthiness leads to the design of a study that is free of bias and that increases the levels of reliability, validity, and objectivity of the research. These measures are specific to the execution of this study.

Rigor and Bracketing

To understand the participants in this study, the researcher set aside known ideas or self-experiences using the *phenomenological mental technique* (Papagiannaki & Shinebourne, 2016). The initial step of this technique, according to Papagiannaki and Shinebourne (2016), is for the researcher to assume the *phenomenological state of mind*. The phenomenological state of mind differs from the usual mentality or methods used for comprehending the world. In the phenomenological disposition, the examination sections are everyday life experiences that depict one's regular learning, which are used to investigate the information. The analyst sets aside personal hypothetical, social, and experiential presuppositions. The concept of *sectioning* originates from Moran's (2009) theory in which the scientist makes him or herself available to the information without

determining its legitimacy. In this context, being available implies that the analyst permits himself or herself to see the information as it emerges organically, without uncertainty or judgment. By adhering to this technique, the researcher can remain in tune with the phenomenological motto “back to the things themselves” (Papagiannaki & Shinebourne, 2016).

Ethical Considerations

Consideration of ethics in research is essential, particularly when it involves human subjects. The principal concern of a researcher should be the safety of participants and respect for their way of life. Thus, the participants’ privacy and confidentiality should receive complete protection (Lorig & Holman, 2003). In this study, protections employed included the informed consent, anonymization of participants, and locked storage of data. The informed consent included a disclosure explaining the study procedures, nature of the study, expected benefits and risks, purpose of the study, and participant compensation. The informed consent was written at a readability level that participants could easily understand (Lorig & Holman, 2003). Participants were informed that the study was voluntary and that they could opt out of the study at any time.

Data from the study is saved in three places: the researcher’s computer, the researcher’s external hard drive, and a locked cabinet that is not accessible to unauthorized personnel (Lorig & Holman, 2003). The documents on the computer and the external hard drive are encrypted and are only accessible by password. The data will be archived for 5 years, after which they will be deleted or destroyed. All data and derived information is anonymous and confidential. The only concern with confidentiality was interaction among participants, which was minimized by individual

interviews, each conducted privately. To the researcher's knowledge, the participants did not meet at any time during the study and did not have previous interaction with one another (Burke, 2011). Once the study was concluded, the researcher did not facilitate any interaction between participants in order to continue to uphold confidentiality and privacy.

The researcher submitted an application for the study to the Institutional Review Board (IRB) of his university, which was approved (Lawrence Livermore National Laboratory [LLNL], 2015). The protocol that was submitted in the application addressed all risks and benefits to participants, including the consent form that allowed participants to make informed decisions regarding their participation in the research (LLNL, 2015).

Role of the Researcher

The role of the researcher in maintaining a professional relationship with study participants is instrumental to ensure an ethical study with minimal bias. It is important to avoid a personal relationship with participants, which can potentially skew results and cause participants to invalidate the data results (Snowball & Willis, 2011). According to Hay-Smith, Brown, Anderson, and Treharne (2016), participants respond to pertinent open-ended questionnaires related to the purpose of the study. The participants should feel comfortable answering interview questions honestly without feeling compromised or judged. The researcher therefore worked to establish a friendly, nonjudgmental rapport with participants to encourage honest responses (Snowball & Willis, 2011).

It is also essential for the researcher to receive and confirm informed consent from all study participants. The informed consent addressed the study background, study procedures, risks and benefits of the study, the voluntary nature of the study,

compensation, whom to contact, and confidentiality in participation and responses (Snowball & Willis, 2011). The informed consent process included an audiotaped oral assent in addition to the hard copy informed consent form. The researcher collected survey responses on the day of the interview. Participant responses were audio recorded and transcribed following completion of data collection. The researcher carefully followed all outlined data collection and confidentiality procedures to avoid invalidation of the data.

Chapter Summary

This chapter described the research methods for this study. The research design was a qualitative phenomenological case study that allowed participants to share their experiences with SCD and its complication of priapism and how they have affected their lives. Participants were adolescent males between the ages of 16 and 19 with a history of SCD and priapism from a young age. The open-ended interview structure was employed to obtain in-depth information regarding the influence of psychosocial state and cultural beliefs regarding SCD on participants. A total of seven adolescent males from the community comprised the sample frame. All information from participants has been anonymized and kept confidential.

CHAPTER 4

FINDINGS

The findings collected for this research study are discussed in this chapter. The data collection process for this study was thorough and closely followed the procedures outlined in Chapter 3. Participant responses were verified for accuracy by interviewees following collection of data. The sample size of seven participants met data saturation, and all participants met the selection criteria of having been diagnosed with SCD and SCD-related priapism. Furthermore, all members of the sample group fell within the required age range for this study, ranging from 16 to 19 years of age. Individuals living with patients meeting these conditions were also considered for inclusion in this study, as it was determined that such individuals would contribute valuable information regarding the experiences of their cohabitants.

Demographics

Demographic data were collected for each participant in the study (see Appendix A). This personal information included age, level of education, household population, ethnic background, religion, medical insurance attributes, and sickle cell condition type. The data were collected from seven respondents, and this demographic information was used to validate the sample size and participant eligibility as presented in Chapter 3.

As stated previously, the objective of this study was to explore the lived experiences of male adolescents with SCD and priapism to better understand the condition and its related complication. The researcher validated that all participants had experienced priapism either directly or from a secondary perspective. Other requirements for inclusion in the study included that participants be male and be in their adolescence

between the ages of 16 and 19. All participants fell within this age range, with 86% of the respondents aged 19 years old and 14% aged 18 years old.

Respondents were also asked to provide their levels of education. The majority of the respondents were in grade 12 at the time of the study, while the rest were distributed between college and high school. At such levels of education, respondents were likely able to reason critically without bias toward questions asked by the researcher.

In terms of race and ethnicity, 100% of participants identified as Black/African American. Although this research study aimed to include persons of heterogeneous backgrounds, the sample population is representative given that the majority of the SCD patient population in the U.S. is African American. The religious affiliation of study participants was also recorded. The majority of participants (57%) identified as non-Protestant Christians, with a smaller subset (14%) identifying specifically with Protestant Christianity. The remaining 29% of respondents identified as non-Christian.

Data regarding participants' medical insurance access was also collected. SCD patients' abilities to receive proper medical care and important medical attention depend upon adequate funding, usually through insurance. The majority of study participants (86%) had medical insurance.

Finally, participants' SCD type was collected as part of the demographic data. Results indicated that 43% of respondents have HbSC and 57% have HbSS.

Findings

The responses collected from semistructured interviews with the study participants are presented in this section. The researcher met individually with each of the seven participants, facilitating semistructured interviews following the Interview Guide

presented in Appendix C. Interviews lasted approximately 1 to 2 hours. The researcher employed additional probing questions when needed to elicit deeper responses from the participants. Participant responses are grouped by interview question, with categories specified for each question to facilitate data coding and analysis.

Interpersonal Relationships

The first question pertained to the frequency and manner in which interviewees interacted with medical professionals. These visits were categorized as either positive or negative. The majority of the respondents regarded their interactions with medical practitioners at these visits as positive due to the usefulness of information provided and/or because they found that these visits supported their overall health either when sick or when attending regular checkups. In contrast, those participants who found the experiences to be displeasing often attributed it to hospital staff seeming apathetic toward them or uninformed about their condition.

However, other participants described their experiences as unsatisfactory due to uncomfortable or stressful events which occurred within a hospital setting. Such emotions may stem from a sense of societal rejection and could even heighten levels of depression if not carefully addressed. One participant's response particularly demonstrates the emotional stress connected with seeking medical care for SCD and priapism:

You know, this, I'm, I'm in a lot of pain, a lot. And it really, it brings me to my knees and bring me to tears. You think that I'm doing whatever it is that I'm doing on purpose, so I can come here? No, it's the last place I want to be.

Interviewees were also asked who helped them the most during periods of illness. These responses help indicate which individuals in these patients' lives understand them

and the challenges they face. Meaningful involvement from another person has been found to leave patients feeling supported, helping to minimize both suffering and depression (Sehlo & Kamfar, 2015). The most common caregivers were found to be respondents' mothers, fathers, siblings, friends, and support groups.

All 7 interviewees stated that their mother helped them the most, followed by 3 stating that their fathers helped them the most in the absence of their mothers. Two other participants stated that, in the absence of their mother and father they had the most help from their siblings. Another 2 participants stated that, in the absence of their mother and father they had the most help from friends. Only 1 participant stated that he had help from his support group. About his mother being the most supportive person, one respondent explained:

I would say my mom really helps me. My mom's probably one of the driving forces in my life when it comes to my, comes to my health and my well-being. She's the one that keeps me on track to, I go to my doctor's appointment. She's helps me keep my appointment and she really reminds me to do that and lets me, helps me keep track of all my medication and when I'm not able to speak, she's able to speak for me.

The apparent poor quality of support offered by friends is worth noting, as many male adolescents spend much of their time engaged with such persons at school and other social activities. Therefore, if the social support offered by friends could be improved, such a relationship may positively affect depression levels, as discussed in the following sections.

Knowledge and Memories of Conditions

Knowledge is a vital component of patients' relationships with their support networks and in terms of the patients' mental health. Increased awareness regarding SCD and priapism may decrease societal stigmatization and discrimination, eventually decreasing patients' levels of depression. Because priapism is associated with SCD, this study asked participants' their knowledge of both the primary disease and its related complication. Knowledge regarding diseases or conditions is often gauged by one's understanding of symptoms or treatments for the conditions, as well as a general understanding of the conditions themselves.

Based on the participants' responses, four categories were developed. The first category of individuals encompasses those with little or no understanding of the condition or complication and who are largely uninformed on the matter. The next category encompasses those who know the basic definitions for their condition or complication and are able to explain them in simple terms. The third category includes those with a more in-depth understanding of their condition or complication who are able to both describe their condition/complication and the associated symptoms. The final group includes those with an excellent understanding of their disease or complication who can describe the associated symptoms and are knowledgeable about current treatment options.

Nearly all respondents had some level of awareness regarding SCD and priapism; only 19% claimed to be uninformed while the majority, 81%, claimed to possess some knowledge of their condition or complication. It was also evident that respondents were largely more informed about SCD than priapism. Such a finding makes sense given that

not all individuals experiencing SCD face the prospect of priapism. However, many respondents expressed an in-depth understanding of both the disease and its complication, which is likely due to their interrelatedness and participants' dual diagnosis with both SCD and priapism.

Participants were also asked to share about their individual experiences. Their responses included sharing memories they had of their first encounter with their condition or complication or during an especially critical situation resulting from their illness. Most of the respondents connected their earliest memories with priapism to an acute episode or traumatic event at a young age.

Several interviewees also associated stress experienced by their mothers with an early memory of priapism. Perhaps reflecting the aforementioned findings that respondents' mothers often offered the most supportive care, participants also demonstrated concern with the manner in which this complication affected their mothers. For example, one participant recalled:

My first experience is, was, uh I was around four, I would say, and I was rushed to the hospital. My mom was in the car driving me to children's hospital, Los Angeles. And, uh because I was wailing, I was crying, I didn't understand the pain and I didn't understand why am I hurting 'em.

The majority of early SCD memories were associated with their occurrence at hospitals. This result was likely due to the fact that patients spent much time in hospitals following the onset of the disease. Many respondents found the hospital setting to be depressing at times since they associated being in the hospital with ill health and injury.

In contrast, some participants first experienced SCD within more positive environments, which can be a vital factor in patient well-being.

Participants' Current Statuses

Because this study is concerned with the psychological effect of priapism resulting from SCD, a review of the participants' current health status in regards to SCD and priapism is also essential. Such knowledge will help inform our understanding of the complication, the challenges patients currently face, and key contributors to possible depression. Participant responses regarding patients' current health statuses were divided between their experiences with SCD without priapism episodes and their experiences with the associated complication of priapism to demonstrate how each one affected participants at the time of the study. For patients experiencing priapism, the majority of respondents felt that it predominantly affected them on a mental level, although some reported being affected physically as well. For patients with SCD without priapism episodes, in contrast, respondents felt equally affected both mentally and physically. A number of participants also explained that SCD affected them at school.

A crucial observation from these data is that both SCD and priapism were found to affect individuals adversely in terms of physical health. However, both mental and physical impact were responded at nearly equal levels, suggesting that the physical effects of both SCD and its complication of priapism had a psychological effect on the patients. In addition, considering the social stigma associated with SCD and priapism (as mentioned by participants), the psychological implications of living with this disease and its associated complication could reach such heights that respondents experienced more

of a mental than physical impact with regard to priapism. One interviewee, when asked how SCD and priapism affected his life, responded:

I think another area that it affects me but not really affects me cause (coughs), just I guess mentally it would affect me in like my, my love life and working with, and dealing with girls. But physically I, you know, I try to, I really just keep my head in the books so I'm not really dealing with any girls. But (coughs) it just makes me think, you know, if I were, would I have an issue, would I have a problem.

Some of these findings may be explained by the fact that the physical limitations experienced by respondents made it less possible for them to leave the home or hospital. With their ease of movement greatly reduced, respondents were not able to engage in as much physical activity as they may have desired. While patients described being able to move from one location to another, they often noted feeling tired as a result.

By examining the environments where patients spend the majority of their time while dealing with the condition and its complication, even more insights into respondents' lives with priapism and SCD are offered. This examination is critical in facilitating more positive environments for care that effectively aid patients and help support a more positive outlook.

For priapism, the majority of interviewees experienced the extreme effects of the complication at home or at a hospital. One interviewee described his experience in the hospital in detail, offering a clearer picture of this location as one of importance in patients' lives:

My current experience, I'm still, still recovering from what I have, in my last experience and discharge from the hospital and going through the, I think it was, I

don't the exact name but I want to say a hyperbaric chamber. I had to go into like a pressurized chamber, so they could work with that and they also had to have, go in and do some type of procedure where they had to remove the blood clot from my penis.

Interviewees were also asked to describe the various elements of their daily experiences. Based upon an analysis of these responses, an average overview of activities was created. The results largely confirm the data collected regarding care needs illustrated earlier in this chapter, with nearly half of the day spent tending to one's health.

In contrast, a mere 27% of the day was spent trying to maintain one's normal school or work routine. The remainder of the day was spent attempting to deal with pain, accounting for 18% of the respondents' average time. This statistic reveals that pain plays a crucial role in the daily life of patients with SCD and priapism.

Lastly, respondents spent 9% of their day, on average, with friends or engaging in social activities. This finding reveals that patients allot minimal time for leisure. While patients may have fewer friends or spend less time socializing as a result of their disease, an analysis of the day-to-day life of respondents reveals that minimal time is spent on activities other than those related to their condition and its complications. Because social engagement generally leads to the creation of a positive environment, such activities often help improve patients' mental well-being and reduce depression. The lack of this type of social relief is evident in the following response from one interviewee:

A normal day (laughs), I think a normal day in the life is just to wake up in pain and just you have to figure out how you're going to deal with that pain. You have to assess what, what is the pain that you're, you're dealing with at this moment and

this day. And how are you going to now combat that, how am I going to, you know, figure out what I want to do with my day? ‘Cause I still, I still have a life, I still have my, my day that I have to go through even though I’m dealing with this pain.

The next interview question explored patients’ most recent experiences with their condition or complication. For many participants, the most recent acute episodes were faced while at home. The majority of respondents had most recently experienced an episode related to priapism; this question, however, must be viewed with the understanding that SCD is directly associated with priapism. Responses suggest that participants noted the effects of priapism more readily.

When asked about the medications that they used at home and while in the hospital to treat SCD and priapism, participants reported using a range of narcotics, painkillers, and natural and alternative remedies. Tylenol and Hydroxyurea were the two most frequently used home medications. It is worth noting that sympathomimetics (Phenylephrine and Sudafed) were only given to 43% of patients for at-home treatment, despite their common usage in treatment protocol for priapism-prone SCD patients. Morphine and IV hydration were the most frequently used treatments while in the hospital.

Effect on Quality of Life

Study participants were also asked how their lifestyles were affected by SCD and priapism. Responses from participants were divided into periods with and without acute priapism episodes. It was found that participants in both circumstances saw their lifestyles affected by their condition or complication; relationships, in particular, were significantly influenced in both groups. This finding builds upon the previous findings;

because living with SCD and priapism minimizes the time spent with friends or social engagement, this aspect of patients' lives is directly affected. This finding is reflected in one interviewee's response:

Well, let's see, how, it, it's sort of, uh, embarrassing, of course, I don't tell people about it. You know, I, I can't go to it and talk to it with my mom, my girlfriend because it's a, a sensitive issue.

Those living with SCD without priapism episodes also found their school lives significantly influenced as a result of living with their condition, likely due to the extra time spent in hospitals and other healthcare facilities. Other patients may require extra monitoring and care, requiring them to utilize adaptive, less traditional education methods, such as homeschooling. Such arrangements likely affected participants' interactions with the larger world while causing some relationships to deteriorate.

When asked about their concerns regarding their disease, participants primarily shared worries about their futures and how their condition or complication will continue to impact their lives. Such anxieties can result in further psychological challenges due to the constant physical suffering they endure on a daily basis. Interviewees largely worried about their bodies' abilities to support the achievement of their future goals. However, concerns were far from limited to this particular anxiety, extending to such arenas as one's ability to produce and care for children, maintain healthy sex lives, and avoid hospitalization.

Participants experiencing priapism episodes revealed that most of their worries were associated with ambitions for the future, with the key worry surrounding the ability to function on a sexual level. Due to the fact that all respondents were at the age when

most individuals begin to explore or discover their sexuality, such a finding is far from surprising. Sexual worries may have also translated into concerns about reproductive capacity. When asked about the prospect of having children in the future, one interviewee said:

Doctors say that there is issues that could happen because of priapism and you won't be able to have an erection and you won't be able to ejaculate because of the constant episodes of priapism. So that's why you got to get to the hospital right away. And I said, I want to have kids, I'm young right now but as I get older I want to maybe have two or three kids.

This patient's concerns reveal a lack of understanding of how priapism affects ejaculation and fertility. Ejaculation is actually not impacted by penile abnormalities like those caused by priapism. Patients that frequently experience priapism episodes are encouraged to have regular sex with their partner or to ejaculate manually in order to maintain normal sperm production. However, priapism does not damage the testes or change hormones and therefore does not affect fertility.

The possibility of future episodes also proved a significant concern, in addition to the experience of pain as a result of priapism. However, a number of participants also shared that they were unconcerned about potential changes or challenges. Such a response may be attributed to the psychological tactic of blocking out worries.

Concerns and worries based on SCD were notably unique compared to those associated with priapism. The most significant concern for SCD patients when they weren't experiencing priapism episodes was the ability to live a healthy life. A number of individuals found the suffering as a result of the vast symptoms associated with SCD to

be a significant hindrance, rendering them incapable of living a normal life. According to interview responses, living with the condition led to a nearly complete transformation in their lifestyles. Following the condition's onset, subjects described needing extra care such as constant monitoring, becoming tired more quickly, and an inability to be exposed to certain environments due to fear of their condition deteriorating. Taken together, these new lifestyle conditions made it difficult for participants to engage in some of their previous passions such as sports and other strenuous physical activities. Their new situations also made maintaining their previous social circles more difficult, often decreasing both their size and quality.

Longevity and health were other major concerns for interviewees. These worries were followed in significance by preoccupations with being trait carriers, future pain, and potential hospitalization.

Individuals with priapism or SCD also have specific needs associated with their condition or complication. Many participants believed that, should these circumstances be met, they would be relieved of much, if not all, psychological and physical discomfort.

Respondents experiencing priapism largely identified self-care related to their complication as their greatest need at the time of the study; notably, this was also the primary concern for those with SCD. However, while the need for pain relief was a concern for respondents with SCD, this was not the case for those experiencing priapism episodes. Similarly, home medication was noted as a current need for persons experiencing priapism while it was not emphasized by those with SCD without priapism episodes. These data illustrate that, for some SCD patients, sickle cell pain is a more significant concern than pain from acute episodes of priapism. For priapism patients, their

primary concerns appear to lie elsewhere and may be associated with a desire for privacy and avoiding social stigma or embarrassment.

Interviewees also shared their challenges surrounding specific tasks and activities in their lives.

The most common difficulty that respondents faced was engaging in recreational activities and sports. Such a finding is likely attributed to the individual symptoms associated with the patients' condition, including shortness of breath, exhaustion, and other physical side effects. For example, one respondent explained:

It's not as, it's not easy growing up with an illness. Especially trying to go to school, I can't finish my classes, I can't uh, I can't concentrate sometimes because I'm in pain in, in school. And I actually have to leave class or drop my classes because I can't deal with this disease and school at the same time.

The next crucial challenge was simply dealing with pain, as noted by 57% of the respondents. Relationships and academics were both recorded at 43%, demonstrating other key areas of difficulty. As further demonstrated in earlier interview responses, social engagement and school life is greatly minimized for patients with SCD and priapism since they are often relegated to the home or healthcare facilities.

Another interview question pertained to the impact of the condition or complication on participants' family members. Perhaps due to the more intimate nature of these settings, the family was found to offer the best supportive care to the patient. Notably, however, while many challenges are faced by the patient alone, their family members also endure much hardship. Respondents noted that family dynamics with different members of the family were often affected as a result of their condition or

complication. This suffering is largely psychological in nature, likely resulting from fear of losing their loved one or witnessing their loved one's physical suffering.

Furthermore, the data revealed that of all family members, the mother is most affected by her loved one's complication in the case of priapism. In contrast, for those patients with SCD not experiencing acute priapism episodes, the family as a combined unit was equally affected. Thus, it appears that, apart from the patient himself, the family is most affected by the condition. This finding is likely due to the fact that the family is closest to the patient's environment and thus witnesses their condition firsthand.

As with family members, a patient's community can have a huge impact on the life of a male adolescent with SCD or priapism. Societal views of SCD and its complication of priapism may affect patients either positively or negatively. Interviewees largely felt that society responded negatively toward individuals with their condition, which may be indicative of various kinds of discrimination or alienation. Participants indicated that the ongoing painful, acute, and chronic events they experienced from SCD can be more stressful than episodes of priapism alone.

Nevertheless, some members of society also offered hope to those living with SCD and priapism, perhaps as the result of different programs mandated to generate awareness regarding the disease and its complications. By providing a positive and helpful environment, such communities can encourage persons living with SCD and priapism and even help decrease patients' depression levels. Society thus has a significant role to play in supporting patients with SCD and priapism.

The effects of priapism tend not to be openly addressed in society due to the sensitive nature of the complication. As such, many patients reported multiple emotional changes as a result of experiencing this complication.

The majority of emotions experienced by participants can be classified as negative in nature. Key amongst these negative changes were feelings of shame, followed by those of fear and stress. These emotions likely stem from the sensitivity regarding the sexual area of the body in which priapism occurs. Because adolescents are just beginning to understand and explore their sexualities, they may be particularly sensitive to discussing this complication. Other negative emotional changes that participants shared include frustration, pain, isolation, and hypersensitivity.

However, respondents also noted positive emotional changes such as increased proactivity, strength, and caution. As a result of their ability to face the complication, respondents interviewed explained that they have become more prepared when facing change and difficulty.

The emotional changes associated with SCD are similarly as negative as those that participants experienced with priapism. However, unlike those associated with priapism, the negative emotions are much more diverse.

Most of these emotions are directly correlated to the physical suffering participants endure with SCD, which participants explain leads to experiences of sadness, depression, and suffering. However, some participants also experienced positive emotions resulting from living with SCD, including more focus, vulnerability, empathy, and strength.

Advice for Others

Participants were also asked to share their advice for other patients living with SCD and priapism. The responses demonstrated the manner in which participants seek to build a community with other patients.

The proactive steps that most participants suggested included talking to others about one's condition or complication and taking charge of one's life. Through talking to others, respondents added, one can relieve much stress. Participants shared that talking to other people allows a patient to distract his mind from negative thoughts that come about when one is idle. One respondent built on this idea of community by explaining:

You know, go to schools or go to, you know, other health facilities, talk to 'em about sickle cell and what, what's going to come with that and sickle cell will get priapism. You know, I think they should have, like, people at schools talking about it. You know, if you're going to be a doctor, well, have them talk to a doc. Let them have a list of the different diseases and the different side effects of those diseases, you know. And what comes with them, yeah.

In terms of hopes for the future of other patients dealing with SCD and priapism, interviewees largely focused on the importance of community, education, and medical services.

Chapter Summary

The results of the interviews conducted as part of this research study demonstrate the significance of various themes associated with living with priapism and SCD. These themes provide a clear understanding of the challenges that these male adolescents face on a daily basis, including those that are both mental and physical in nature.

Analyzing these challenges revealed the role that society plays in terms of patients' psychological health. According to respondents, society did little to provide the support required by patients living with SCD and its complication of priapism. Instead, the family proved to be the key support system for participants; thus, both the private nature of the condition and its complication and patients' reliance on the intimacy of familial relationships was shown to make it difficult for patients to open up to others. Furthermore, normal activities such as school and social activities were shown to be negatively affected, further isolating patients.

However, these responses also demonstrate the dreams and hopes that those living with SCD and priapism continue to hold. In the following chapter, the findings of this study will be discussed in relation to extant literature, and conclusions and recommendations will be presented.

CHAPTER 5

DISCUSSION

The discussion, recommendations, and conclusions for this qualitative phenomenological case study are presented in this chapter. The results of this research study were presented in Chapter 4, including details regarding the participant demographics and data collection and analysis procedures. There were a total of seven participants in this study, all of whom met the selection criteria of being male, being diagnosed with SCD and priapism, and being aged between 16 to 19 years of age. This study was guided by three research questions:

1. What are the experiences of priapism among male adolescents with SCD?
2. How has priapism affected the lives of male adolescents with SCD?
3. What is the impact of the male adolescent's daily experience with priapism and SCD on the family?

This chapter first presents a discussion of the study findings in relation to previous literature, followed by a consideration of the implications for practice and the limitations of the study. Finally, recommendations for future research are provided, as well as concluding thoughts on the study.

Discussion of Findings

The findings of this study both confirm some previous literature and lend further insight that helps expand researchers' knowledge regarding the lived experiences of individuals with SCD and priapism. These findings are organized by theme and discussed below in relation to extant literature.

Concerns with Sexual Performance, Stigma, and Associated Psychological Effects

The male adolescent participants in this study who were suffering from priapism stated that one of their greatest fears was a lack of sexual performance, including concerns with their ability to function sexually and their future reproductive abilities. Their fears are partially valid given that research indicates that priapism can lead to impotence, or the inability to get an erection, if left untreated for too long (Joice et al., 2015). However, priapism does not cause infertility, and many participants seemed to be misinformed about this fact.

Researchers have also found that individuals with SCD often experience delays in puberty (Joice et al., 2015), which can have a negative psychological impact on these adolescents as they face delayed sexual development (Sehlo & Kamfar, 2015). The present research study found similar psychological effects associated with priapism and participants' fears surrounding the long-term sexual impact of this complication. One participant expressed a desire to have children in the future and explained that he knew that he needed to seek treatment right away during a priapism episode in order to avoid long-term complications with his reproductive health. His concerns were somewhat misguided and pointed to a lack of accurate information regarding priapism and its effect on reproductive health. These findings related to patients' concerns are in line with previous literature, but they provide further depth regarding priapism individuals' lived experiences and the psychological impact of the sexual aspect of the complication.

Previous research has also found that adolescents with priapism often experience stigmatization and isolation as a result of their complication, particularly when it comes to recreational activities (Sehlo & Kamfar, 2015). Participants in this study reported

spending only 9% of their time at social gatherings with friends or engaging in recreational activities. The physical symptoms and side effects of this complication influenced interviewees' ability to engage actively in recreational activities. One participant reported struggling to participate in school activities due to high levels of pain and often having to abandon those activities. However, other participants were not willing to participate in such activities due to feelings of discrimination or stigmatization from others. Previous research has found that such feelings of expected discrimination and internalized stigmatization are relatively common among adolescents with SCD and SCD-related priapism (Bediako et al., 2016).

One of the most pressing current needs that priapism respondents in the current study reported was self-care related to their health, which they described as a desire for privacy as well as avoiding social stigma or embarrassment. These participants preferred to stay away from other people and thus refrained from participating in social or recreational activities. Therefore, while the results of the current study confirm the findings of Sehlo and Kamfar (2015) and Bediako et al. (2016), they also demonstrate the complexity of this complication and the combination of both physical and psychological factors that are involved in the limited recreational activity of individuals with priapism, which could be associated with physical pain as well as feelings of isolation and stigmatization.

Quality of Life and Support Systems

The quality of life and lifestyles of participants in this study were also greatly affected by SCD and its complication of priapism. Participants with and without acute episodes of priapism found their relationships with family and friends and their school

participation significantly affected. The deteriorated relationships and impacted participation were likely due to the amount of time spent away from these environments, including in hospital care or at home recovering from a traumatic health episode.

Despite the condition's impact on their relationships, however, participants still placed a high level of importance on support systems to help them cope with their health conditions. Previous research found that social support systems are impactful for individuals diagnosed with SCD or who are experiencing priapism episodes, as those with such social support will have fewer adjustment problems (Sehlo & Kamfar, 2015; Telfair & Gardner, 1999). Family was the key support system for participants in this study, with mothers accounting for the majority of responses and fathers coming in second in terms of the patients' primary support system. Friends accounted for minimal responses, which is slightly concerning given that male adolescents tend to spend a large amount of their time with friends or at school. However, this finding is a possible consequence of the social isolation that many of these individuals reported experiencing and is in line with previous research on stigmatization and isolation as a result of SCD and priapism. Considering the intimate nature of priapism, participants appeared to feel more comfortable relying on family members for support and assistance when it came to their health issues. This study's findings regarding the types of support systems on which SCD and priapism patients rely contribute new information to research in this area, expanding the findings of Sehlo and Kamfar (2015).

Experiences With Medical Care

Participants in this study reported mixed experiences with medical care, whether it was direct interactions with medical practitioners or experiences with hospital care.

While the majority of participants regarded their interactions with health care providers as positive during medical visits, some participants had negative experiences and reported that hospital or clinic staff acted apathetic or were uninformed about their condition or complication. Several participants recalled stressful or uncomfortable events occurring while at the hospital, with one respondent remembering being brought to tears because of the shame he experienced during a hospital visit.

The negative experiences reported by some participants in this study confirm previous research findings. Researchers found increased stigma around SCD among medical practitioners, where patients diagnosed with the disease were referred for medical care more frequently simply because they had the disease (Bediako et al., 2016). Other researchers found that practitioners treated patients with SCD and other chronic pain diseases nontherapeutically (Adegbola, 2011) and regarded SCD patients as generally difficult and challenging to communicate with (Thomas, 2006). These types of stigma-informed attitudes among some medical practitioners appear to be represented in the experiences of some of this study's participants. However, it is promising that more participants had positive than negative associations with medical visits overall. These respondents reported being able to access relevant information regarding their condition or complication and receive helpful updates on their health status during their medical checkups.

Study participants' preferences for self-care and home medication when it comes to their health are in line with previous findings as well. Furtado et al. (2012) reported that priapism patients were likely to delay seeking medical attention because of the stigma associated with the complication. The participants' responses in the present study

reflect a similar preference for remaining isolated and seeking home treatment whenever possible. In regard to managing a personal crisis or episode with their health, participants with priapism reported being proactive and using self-care and home medication to alleviate the acute episode. These findings appear to align with Furtado et al.'s (2012) research, although the emphasis on self-care in the current study is a new finding that could be investigated further.

Knowledge of the Condition and Advice for Others

Previous research has suggested that improved awareness of SCD and priapism may significantly decrease instances of discrimination and stigmatization against individuals with these health issues (Thomas, 2006). However, previous literature has not examined individual patients' knowledge of the disease since the focus of such research has mainly been on the types of situations they encounter and how they manage the condition. This study was therefore the first that the researcher is aware of that asked the interviewees how conversant they were with their condition or complication. The goal was to gauge a participant's understanding of the disease based on his comprehension of the symptoms and treatments.

This study found that participants were relatively well informed and aware of SCD and/or priapism. The findings show that 81% of the respondents had some knowledge about priapism, while only 19% claimed to be uninformed about priapism. This finding is telling given that not all SCD patients experience priapism, so it appears that those who do not experience priapism frequently may be less informed about it. This points to the need to further educate both the broader public and the SCD patient

population about priapism in order to help avoid stigmatization and isolation for priapism patients.

Finally, the study participants shared advice that they had for other individuals suffering from SCD and priapism. Participants advised talking to others about the condition or complication in order to relieve stress and distract them from any idle thoughts. They also recommended taking care of one's overall health and seeking the support of their family and community. These insights lend further depth to the existing research on the lived experiences of individuals with SCD and priapism, offering several areas for improving practice and conducting further research. These areas are explored later in this chapter.

Implications for Practice

Sickle cell disease and its associated complication of priapism are rarely discussed in our broader society. Therefore, this research study intended to shed light on a disorder and its complication which are already characterized by isolation and discrimination. The findings gathered in this study demonstrate the many gaps that currently exist in the healthcare system in terms of adequately supporting adolescent male SCD and priapism patients. Several recommendations for practice are presented in what follows.

Given the varying levels of knowledge regarding priapism among participants, it is recommended that awareness programs or trainings be developed around both SCD and priapism within our healthcare system. This recommendation is in line with that of Thomas (2006), who recommended cultural awareness training in an attempt to increase the general public's knowledge and awareness of SCD and thus alleviate some of the

stigma surrounding the disease. This study's findings indicate that there is still a need for such trainings, both for healthcare practitioners as well as for the broader public. In addition, a program that educates SCD patients specifically regarding priapism as a complication associated with the disease is advisable for preventative purposes and in order for current SCD patients to better support their peers who may be dealing with priapism. It may be useful to incorporate such an awareness training into current SCD support groups or outreach programs.

Given the fact that the parents or guardians of SCD and priapism patients were also included in this study, it is recommended that further training and support be made available to the families of these patients. According to the research findings, parents or guardians were the most significant support system of individuals suffering from both SCD and priapism, and it is therefore important that they are well informed about their child's condition and that they are offered psychological support themselves to help them cope with caring for their child. Developing support groups or other sources of support would allow parents or caregivers the ability to share their concerns and worries with others and to find comfort and validation in the experiences of other parents in the same situation.

Additionally, hospital intake for patients with priapism should be set up in such a way that it accommodates the intimate and compromising nature of this complication. According to the interviewees, those with priapism were always looking for alternative referrals outside of a hospital or clinic where they could get their medication and seek treatment in private. The majority of individuals only went to the hospital when their situation became dire and they needed immediate medical attention (Furtado et al., 2012).

Several participants complained about the poor treatment they received in the hospital and the lack of awareness among hospital staff about SCD and priapism. Therefore, it is strongly recommended that procedures be put into place for such patients to feel more comfortable when entering the hospital setting in order to avoid further health complications or psychological impacts stemming from avoiding going to the hospital.

Finally, it is recommended that additional support groups be offered for patients with priapism and SCD. Since participants' experiences of the condition and its complication vary widely from individual to individual, receiving generic medical advice from their healthcare provider will likely be insufficient to help individuals cope with all aspects of SCD and priapism. Therefore, being able to share their experiences with others in the same situation and hear about others' different experiences provides an additional level of support and source of information for this population. These types of groups are also a means through which issues of anxiety, discrimination, and isolation can be addressed and potentially alleviated.

Limitations of the Study

This qualitative phenomenological case study utilized semistructured interviews to obtain information about the participants' lived experiences with SCD and priapism. However, there are some limitations associated with the study, which encompass sample size, location, time limitations, and interview setting.

Given the qualitative design and limited sample size used in this study, the findings are not generalizable to the broader population (Schumacher, 2010). The researcher's location was another limitation of the study. Since the sample population was only drawn from the southern California region, the findings were not representative

of the broader national population. In addition, the study findings are not representative of all SCD patients since the sample population only included male adolescents. This research study was therefore not able to examine the effects that priapism or SCD have on the general male population.

Time was also a factor given the time limitations associated with conducting doctoral research of this nature. Given the finite amount of time allotted for dissertation research, the duration of the study was required to be quite short. While a longitudinal or longer-term study would have been of interest to the researcher in order to follow the participants into adulthood, the strict parameters of dissertation research did not make this possible.

The challenge of finding a suitable location in which to conduct participant interviews was another limitation of the study. While the researcher made every attempt to locate suitable settings in which to conduct each interview, the locations were not always deemed adequate by the participants themselves. Certain participants might find a location acceptable while others reported that it made them uncomfortable. Since some participants felt uncomfortable with the interview setting, they may have withheld valuable information that could have contributed further to the study. Nevertheless, every attempt was made by the researcher to put them at ease during the interview process.

Recommendations for Future Research

Given that SCD-related priapism remains an understudied complication, there are countless potential areas for future research, ranging from the causes of the complication, to its prevalence in male adolescents, to the most common forms of priapism, to medical advancements and new types of medication to alleviate pain associated with the

complication. However, the recommendations given in this section are specifically related to the findings of the present study and future research directions that could expand on this study.

It is recommended that future research explore the issues of stigmatization and isolation in more detail. The time limitations of the present study made it difficult for the researcher to exhaustively explore the reasons why patients suffering from priapism prefer to be in isolation. While some of the respondents stated that they perceived that people around them were judging them and they thus decided to separate themselves, a different or prolonged study design might yield additional insights into this issue that will expand on the current findings.

It is also recommended that future research expand the sample population to include men of all ages living with priapism. Including both adolescents and older males in a single study will yield information regarding the experience of aging with the complication and the severity of the acute episodes as patients reach different life stages. Such an analysis will be critical in determining the continuous progression of priapism as one advances in age. Respondents of different ages could also provide details regarding the medications and lifestyle that has worked for them, which might prove extremely informative for ongoing treatment of both priapism and SCD.

Finally, it is recommended that future research explore potential ways of increasing societal support for individuals with priapism. This research might include participants without priapism in the sample population and test different types of trainings or educational programs to see which are effective in improving their understanding of SCD and priapism. The study might also facilitate interactions between

patients with SCD and priapism and the general population, including having the general public participate in or sit in on SCD or priapism support groups to determine what effect that would have.

Conclusion

The condition of SCD and its complication of priapism are little known among the broader American public. This qualitative phenomenological case study attempted to contribute to the existing research about SCD patients with priapism episodes by exploring the lived experiences of male adolescents with SCD and the complication of priapism. The findings confirmed much of the previous literature and contributed additional insights regarding living with these conditions as a young male. Living with priapism and SCD impacts the quality of life of these individuals, causing them to spend less time with friends and participate less in school, and compromising their interpersonal relationships with friends and family. However, support systems play a significant role in their ability to cope with the condition and its complication, with family in the primary support role. Moreover, patients' experiences with medical care are varied; while some reported being pleased with the health information and updates they received, others recalled extremely traumatic and uncomfortable experiences in a hospital setting. Both physical pain and fear of discrimination caused the majority of participants to isolate themselves socially and refrain from participating in recreational activities. For similar reasons, many participants avoided hospital treatment and sought alternative care, self-care, and home medication instead.

Based on these findings, the implications for practice and opportunities for future research are broad and varied, as has been discussed in this chapter. It is imperative to

develop awareness trainings or educational programs to better inform medical practitioners and SCD patients about priapism. Moreover, expanding on this research in order to include a larger and more diverse sample population would likely lend further insights that would contribute more information to the current findings.

In addition to the above recommendations, the researcher would like to conclude with a call for additional research into the causes of SCD-related priapism among male adolescents. It remains unclear why this complication specifically affects the male adolescent population, and further research is needed to gain a better understanding of its prevalence. Future studies should analyze whether priapism is hereditary, lifestyle-related, or can affect anyone. Although research has shown that SCD is an inherited disorder, it is unclear whether its complication priapism, which occurs in some but not all SCD patients, might have genetic determinants. Understanding the causes of this particular SCD-associated complication could have a profound impact on the treatment and prevention of such a painful and uncomfortable experience as acute priapism episodes.

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APPENDIX A

DEMOGRAPHIC DATA QUESTIONNAIRE

Date of Interview _____

Age

16

17

18

19

Highest grade completed _____

How many people live in your household?

1 to 2

3 to 4

5 to 6

7 or more

What is your ethnic group?

Black/African American

Mexican American

White/Caucasian

Other (Specify) _____

Decline to state

What is your religious group?

Catholic

- Jewish
- Protestant
- Other (Specify)_____

Have you ever experienced priapism? _____

Type of your Sickle Cell Disease

- HbSS
- HbSC
- HbS beta thalassemia
- HbSD
- HbSE
- HbSO

APPENDIX B

CONSENT FORM

University of San Diego

Institutional Review Board

Research Participant Consent Form

For the research study entitled:

Priapism among male adolescent with Sickle Cell Disease

I. Purpose of the research study

Prince Ocansey is a student in the Hahn School of Nursing and Health Science at the University of San Diego. You are invited to participate in a research study he is conducting. The purpose of this research study is to use a lived experience to understand priapism among male adolescents with sickle cell disease.

II. What you will be asked to do

If you decide to be in this study, you will be asked to:

- Complete a ten question demographic questionnaire that ask you questions about your age, ethnicity, highest grade completed, employment status, household yearly income, type of your sickle cell and your religious group.
- Participate in a private face-to-face semi-structured interview about your lived experience with sickle cell disease and priapism
- You will be audiotaped/videotaped during the interview
- Your participation in this study will take a total of 60 minutes

III. Foreseeable risks or discomforts

Sometimes when people are asked to think about their feelings, they feel sad or anxious. If you would like to talk to someone about your feelings at any time, you can call 24 hours a day:

San Diego Mental Health Hotline at 1-800-479-3339

Los Angeles County Department of Mental Health Hotline at 1-800-854-7771

Riverside County Mental Health Helpline at 1-951-686-4357

San Bernardino County Behavioral Health Crisis Hotline at 1-909-386-8256

IV. Benefits

While there may be no direct benefit to you from participating in this study, the indirect benefit of participating will be knowing that you helped researchers better understand the lived experience of priapism among male adolescents with sickle cell disease.

V. Confidentiality

Any information provided and/or identifying records will remain confidential and kept in a locked file and/or password-protected computer file in the researcher's office for a minimum of five years. All data collected from you will be coded with a number or pseudonym (fake name). Your real name will not be used. The results of this research project may be made public and information quoted in professional journals and meetings, but information from this study will only be reported as a group, and not individually.

VI. Compensation

If you participate in the study, the researcher will personally give you \$50.00 Visa card.

You will receive this compensation even if you decide not to complete the entire interview session.

VII. Voluntary Nature of this Research

Participation in this study is entirely voluntary. You do not have to do this, and you can refuse to answer any question or quit at any time. Deciding not to participate or not answering any of the questions will have no effect on any benefits you're entitled to, like your health care, or your employment or grades. **You can withdraw from this study at any time without penalty.**

VIII. Contact Information

If you have any questions about this research, you may contact either:

1) Prince Ocansey

Email: pocansey@sandiego.edu

Phone: (951) 235-3755 or (951) 222-2277

2) Dr. Jane Georges

Email: jgeorges@saandiego.edu

Phone: (619) 260-4566

I have read and understand this form, and consent to the research it describes to me. I have received a copy of this consent form for my records.

Signature of Participant

Date

Name of Participant (**Printed**)

Signature of Investigator

Date

APPENDIX C

INTERVIEW GUIDE

1. Tell me about your visits to your doctor(s).
2. During periods of illnesses, who helps you the most often? Your mother, father, friend, colleague, siblings?
3. Please describe what you know about priapism and sickle cell disease.
4. What do you remember about your earliest episodes of having sickle cell disease?
5. How has your experience with priapism and sickle cell disease affected you?
6. How was your current experience with the sickle cell disease and priapism?
7. What is a normal day for you as you live with sickle cell disease?
8. Tell me about your most recent crisis.
9. What medications help to manage your pain at home?
10. What type of medications are used during hospitalization to manage your pain?
11. How is your life affected by this crisis in terms of work and relationships?
12. Please describe how you personally manage these crisis?
13. What are primary issues of concern about having sickle cell disease?
14. What are your needs related to sickle cell disease?
15. Does sickle cell disease keep you from doing things that you would like to do? What do you do about that?
16. Please describe how sickle cell disease affected your family dynamics?
17. Would you like to tell me anything else about your condition?
18. How is society doing with regard to sickle cell disease?
19. Tell me about your experience with priapism.
20. What emotional changes you have experienced?
21. Tell me about your experience with blood transfusion, if any.
22. Do you have any advice for others with sickle cell disease, their families, and the medical personnel who help people with the disease?

APPENDIX D

LETTER SEEKING SUPPORT FOR THE STUDY

Prince Ocansey
University of San Diego
Hahn School of Nursing
5998 Alcalá Park
San Diego, CA. 92110

Date: November 26, 2016

Sheila Marchbanks
Sickle Cell Support Group
PossAbilities Office
Professional Plaza – Building A
25455 Loma Linda Drive, Suite 109A
Loma Linda, CA 92354

Dear Ms. Marchbanks

My name is Prince Ocansey and I am a third-year Doctor of Philosophy (PhD) student at University of San Diego. I am conducting a qualitative study on the lived experience of priapism amongst male adolescents with sickle cell. I will be conducting a semi-structured face to face interview and will need participants within the 16 -19 age range.

The purpose of this study is to use a lived experience to understand priapism among male adolescents with sickle cell disease. This will help me to understand the kind of life that sickle cell disease brings to an individual as it will focus on perceptions of priapism. It will also provide a sense of direction to health care providers who seek to develop methods of assessing and managing priapism within the adolescent population with sickle cell disease.

For this project, I plan to interview approximately 10 male adolescents. This will be absolutely voluntary. At the end of the interview, each participant will receive a \$30 gift card for appreciation and compensation.

Confidentiality will be maintained throughout the interview. All names and data collected will be stored in a safe deposit and only accessed by my chair Dr. Jan Georges of University of San Diego and myself.

Thank you for your support towards my study. Kindly contact me on 951-235-3755 for any further information or queries.

Sincerely,

APPENDIX E
RECRUITMENT FLYER

Participant are needed in a Research Study:

Priapism among Adolescents with Sickle Cell disease

I am seeking male between ages 16 and 19 years who have been diagnosed with sickle cell disease and priapism. I am a Doctoral nursing student at the University of San Diego conducting a study to look at lived experience of male adolescents with sickle cell disease diagnosed with priapism. Participation involves filling out a form and a face-to-face semi structure interview that takes about an hour. Participants will receive a \$50.00 Visa card. Please contact Prince Ocansey at (951) 235-3755 or (951) 222-2277 for more information or email pocansey@sandiego.edu.