

**SCHOOL OF PUBLIC HEALTH
COLLEGE OF HEALTH SCIENCES
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**QUALITY OF LIFE OF ADOLESCENTS WITH SICKLE CELL DISEASE
ATTENDING THE GHANA INSTITUTE OF CLINICAL GENETICS, KORLE BU
TEACHING HOSPITAL.**

**BY
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DECLARATION

I, Mary Fynn, declare that this thesis titled ‘Quality of Life of Adolescents with Sickle Cell Disease attending the Ghana Institute of Clinical Genetics, Korle Bu Teaching Hospital’ is my own original work as a student of School of Public Health, University of Ghana under supervision. This thesis has never been submitted in part or whole for the award of a degree in any university or higher institution of learning. All references made to other people's work have been duly acknowledged.

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MARY FYNN

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PROFESSOR PHILIP BABA ADONGO

Academic Supervisor

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DATE

DEDICATION

I dedicate this work to all adolescents with Sickle Cell Disease. I know your struggles. It is well.

ACKNOWLEDGEMENT

This thesis has not been produced by sole effort. There are several people worth mentioning who assisted in diverse ways for this dissertation to become a reality.

My very first thanks goes to the living God that I serve, in whom I live, and move, and have my being. I have come this far, because He has made it so.

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Special thanks go to my friends who kept me on my toes, and became ‘home supervisors’, making sure I met deadlines, and encouraging me in the low times.

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ABSTRACT

Background: Sickle Cell Disease (SCD) describes several genetic conditions with the presence of an abnormal haemoglobin S (HbS) in the red blood cells. The commonest types of Sickle Cell Disease are sickle cell anaemia (SS), sickle cell haemoglobin C disease (SC), sickle beta-plus thalassemia, and sickle beta-zero thalassemia. The abnormal haemoglobin causes red blood cells to assume a sickle shape. Large numbers of sickled red blood cells collect, hindering blood flow, leading to vaso-occlusive pain concentrations at the joints. Patients also suffer from persistent haemolytic anaemia, lung and heart-related complications, growth and puberty complications, as well as dysfunctions in organs such as the liver, and kidney. The adolescence period is a stage of transition from childhood to adulthood, marked by a sense of independence, and identity. Quality of life (QOL), a social construct measuring the overall wellbeing of a person, has been shown to be lower in adolescents with SCD, due to the effects of the disease.

General Aim: The study objective was to determine quality of life of adolescents attending Sickle Cell clinic at the Ghana Institute of Clinical Genetics, Korle Bu.

Methods: This research work employed the qualitative approach, using phenomenological technique. In-depth interviews were conducted with 15 adolescents with Sickle Cell Disease attending Sickle Cell Clinic at the Institute, which were audio recorded and transcribed. Thematic analysis was done by coding and recoding in order to generate themes for data analysis.

Results: The outcome of the study is that quality of life of adolescents with Sickle Cell Disease, based on their subjective meanings, and interpretations they assign to their health at this stage of life is good. The findings of this study include the general, emotional, physical, and social and cultural health and well-being of the adolescent patients, as perceived by them. Social factors such as family support and clinical visits were found to have a significant impact on the

quality of life of adolescents with SCD. Culture was found to have no effect. There were no differences between the quality of life of male and female adolescents with SCD.

Conclusion: Generally, quality of life of the adolescents was found to be good, as perceived by them, however, emotional health was found to be lacking in some younger female adolescents. The social factors that are positively associated with QOL are to be encouraged. It is recommended as well that the emotional health of younger adolescents, especially female is considered, and given more attention in care of SCD.

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LIST OF ABBREVIATIONS

GICG	Ghana Institute of Clinical Genetics
KBTH	Korle- Bu Teaching Hospital
QOL	Quality of Life
SCD	Sickle Cell Disease
WHO	World Health Organisation

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DEFINITION OF TERMS

Sickle cell disease- An autosomal haemoglobinopathy disorder characterized by structural aberration of the haemoglobin of red blood cells.

Vaso-occlusive crisis- Painful episodes as a result of blood vessel occlusion by sickled red blood cells.

Adolescent stage: A transition period between the ages of 10-19 years marked by an increase in sense of responsibility as well as independence. For the purposes of this study, adolescence is a period between 13-19 years.

CHAPTER ONE

INTRODUCTION

1.0 Background to the study

Sickle Cell Disease (SCD) describes several genetic conditions with the presence of an abnormal haemoglobin S (HbS) in the red blood cells. The commonest types of Sickle Cell Disease are sickle cell anaemia (SS), sickle cell haemoglobin C disease (SC), sickle beta-plus thalassemia, and sickle beta-zero thalassemia (Lim, 2009). The abnormal haemoglobin causes red blood cells to assume a sickle shape (Edwin, Edwin, & Etwire, 2011). Large numbers of sickled red blood cells collect, hindering blood flow, leading to vaso-occlusive pain concentrations at the joints. Patients also suffer from persistent haemolytic anaemia, lung and heart-related complications, growth and puberty complications, as well as dysfunctions in organs such as the liver, and kidney (Fuggle, Shand, Gill, & Davies, 1996). Records from the World Health Organisation indicate that Sickle Cell Disease affects 5% of the world's population, and has an annual birth rate of 300,000 babies (WHO, 2006).

The disease has been linked to individuals of African descent, and some Hispanic and Asian ancestry as well (Smith, 2016). Smith (2016), reported that Nigeria currently, holds the highest prevalence of the disease with about 150, 000 new births each year. This researcher records that the life expectancy of persons with SCD is relatively low at 50 years of age. However, proper management of the disease could reduce complications, and prolong life.

In Ghana, about 2% of the population has the disease, with a prevalence of 30% forming the carrier population (Edwin *et al.*, 2011). Due to advances and improvements in technology and medical advances, improvements have been made to increase life expectancy, but not as much

in developing countries (Kwarteng-Siaw, Paintsil, Toboh, Owusu-Ansah, & Green, 2017). Kwarteng-Siaw *et al.* (2017) explain that more children survive from the childhood to adolescent stage, and thus there is the need to make provision for their care and facilitate survival to adulthood.

The adolescent stage is a period of transition marked by an increase in sense of responsibility as well as independence (Smith, 2016). Poor disease management strategies, including non-adherence to medication regimes, and others, could amplify the negative effects of the disease. Despite the different challenges that the adolescent faces at this stage, including the physical, social and emotional challenges at this stage which are in themselves compounded and further affected by the presence of SCD, the adolescent as well has to deal with responsibility of the disease (Forrester, Barton-Gooden, Pitter, & Lindo, 2015).

Technological advances have seen improvement in quality of life, however, a study found that health-related quality of life for adolescents was still generally lower as compared with those of their non-SCD counterparts (Dale, Cochran, Roy, Jernigan, & Buchanan, 2011).

Kwarteng-Siaw *et al.* (2017) in a study on transition readiness of SCD adolescents in Ghana concluded that planning for management of the disease at this stage of life should be considered. Therefore, there is the need to assess the quality of life of adolescents at this stage of transition since it has its own peculiar complexities that could compound the challenges of the disease; and to identify the coping mechanisms to deal with such a chronic illness.

1.1. Problem Statement

Globally, SCD has been recognised as a genetic disorder of public health concern, with majority of the sufferers being of African descent especially, and Hispanic descent (Piel, Hay,

Gupta, Weatherall, & Williams, 2013). World Health Organization asserts that the prevalence of the disease is five percent the world over (Edwin *et al.*, 2011). SCD originates from sub-Saharan Africa and is most prevalent here, where malaria is endemic. (Anie *et al.*, 2010; Edwin *et al.*, 2011). Nigeria records the highest statistical prevalence, with about 20-30% of the population having the disease (WHO, 2011). In Ghana, 1 in 3 Ghanaians have the haemoglobin S and/or C gene, and 2% of newly born Ghanaians have the disease. This puts the current state of the nation with regards to SCD at 15,000 babies annually, and 30% carrier-population nationwide, a note-worthy public health burden (Edwin *et al.*, 2011).

With great advances in technological and medical knowledge, quality of life of the sufferers and care givers, and family members, as well as prognosis of the disease has improved (Edwin *et al.*, 2011). However, according to studies from Bakare (2016), stigmatization, discrimination and social isolation are some psychosocial difficulties faced by SCD patients, and their relations.

The adolescence period of development is marked with many problems, and is the time when people are most adventurous and are likely to begin experimenting with risky behaviours that can affect their health in the future (Asnani *et al.*, 2014). Dealing with a chronic illness such as SCD which results in frequent hospital visitations, and prolonged hospitalizations leading to school absenteeism would further exacerbate the physical, and other challenges of this stage.

The purpose of this study is to determine the quality of life of adolescents with SCD, and consider their responses to psychosocial pressures at this stage of development as persons with a chronic illness.

1.2. Significance of Study

Quality of life has been generally accepted as a multi-dimensional construct, useful for the use of evaluating the effects of a chronic disease on the life of an individual (Fuggle, Shand, Gill, & Davies, 1996; Ola, Yates, & Dyson, 2016). SCD is a chronic illness, requiring application of certain practices and lifestyle adaptations to curb the severity of the disease and avoid crisis, and/or early death.

Adolescent stage is marked by the formation of interpersonal relationships, and peer support which actually tends to affect their development. In order to fit in with their cohorts, adolescents with SCD may engage in risky behaviours, such as smoking, drinking hard liquor, and having unprotected sex, behaviours which can lead to depression, and negative self-esteem, and by extension, exacerbate the symptoms of the illness (Forrester *et al.*, 2015), and in effect affect the various domains of the life of the individual. Evidence has shown that SCD adolescents in Jamaica engage in risky behaviours such as smoking, and having unprotected sex (Asnani *et al.*, 2014)

This study seeks to influence already existing policy concerning care of SCD adolescents, considering all other aspects of their quality of life, including physical and psychosocial functioning, and not only considering biomedical care.

1.3. General Objective

To determine the quality of life of adolescents with SCD attending the Ghana Institute of Clinical Genetics, Korle Bu Teaching Hospital.

1.3.1. Specific Objectives

1. To examine the perceptions of adolescents with SCD on how their quality of life is affected by the presence of SCD.
2. To examine social-cultural factors associated with the quality of life of adolescents with SCD.
3. To assess the differences between the QOL of male and female adolescents with SCD as perceived by them

1.3.2. Research Questions

The research questions that will be answered in this study are:

1. What is the QOL of adolescents with SCD?
2. What are the perceptions of adolescents with SCD on how the disease affects their quality of life?
3. What are the social-cultural factors associated with the quality of life of adolescents with SCD?
4. Are there any differences between the perceptions of male and female adolescents with SCD on their quality of life?

CHAPTER TWO

LITERATURE REVIEW AND CONCEPTUAL FRAMEWORK

2.0 Introduction

This chapter presents detailed insight into previous literature and existing studies that have been done on sickle cell disease, its prevalence and clinical variability, the stage of adolescence and its characteristics, as well as quality of life. It also presents a conceptual and theoretical framework that serves as the foundation of the research, as well as information that serves as a basis for discussing the finding of this study. Section one talks about the prevalence of sickle cell disease on a global scale, as well as its history, and epidemiological distribution. Section two talks about the clinical variability of SCD, touching on general signs of the disease among sufferers. Section three discusses the adolescent period as a stage of transition from childhood to adulthood, as well as behaviours related to this stage. Section four reviews existing literature on the adolescent behaviour and management of SCD condition. Section five explains quality of life, with regards to adolescents, and reviews literature on the specific objectives of the study. Section six discusses the theoretical and conceptual framework of the study.

2.1 Prevalence of Sickle Cell Disease

SCD is an inherited disorder of the red blood cells. An abnormal haemoglobin known as sickle haemoglobin present in red blood cells which are used for carrying oxygen throughout the body, has a sickling shape in people with SCD, causing the red blood cells to assume same shape, and thus, the name. The sickled red blood cells die, causing anaemia, and cluster together in small blood vessels, blocking the flow through the vessels and obstruction of the flow of oxygen through the body producing pain, damage to tissues, and severe anaemia of the cells (Inati-Khoriaty, 2008).

The three commonest types of sickle cell disease are; haemoglobin SS or sickle cell anaemia, haemoglobin SC and Haemoglobin sickle beta thalassaemia. Sickle cell anaemia and sickle cell disease are used interchangeably, but are not the same, with sickle cell anaemia being the most severe form of sickle cell disease resulting from the production of only abnormal sickle haemoglobin (Inati-Khoriaty, 2008).

SCD has originally been known to exist in tropical and sub-tropical areas of the world. In recent years, it has been recognized by the World Health Organisation (WHO) as a disease of public health burden worldwide. SCD records about 300, 000 births annually, most of which are found in low- and middle-income countries, and especially in Africa. It has been recorded as a chronic disease, and hence its ill effects on global healthcare as a burden have been noted. SCD is prevalent in African, Hispanic and other Mediterranean areas and populations, with the disease recording high prevalence in Africa due to the advantage of the sickling status against malaria (Anie, Egunjobi, & Akinyanju, 2010).

However, the disease has been found to spread to other areas of the world due to emigration and immigration of people for various reasons. Red blood cells become sickled in the disorder, causing a blockage of small vessels in the blood, resulting in severe pain episodes, anaemia, and some organ damage. Simple steps including early infant diagnosis, accessibility to hospital treatment, and proper drug administration could lead to better and longer quality of life (Modell & Darlison, 2008).

According to Smith (2016), proper management of the disease can reduce the risk of mortality, as well as the complications that arise from having the chronic condition. Bone marrow transplant offers some form of cure to children with SCD with high event-free survival rates,

however, it is a risky and difficult procedure (Smith, 2016) involving donation of an identical human leukocyte antigen from a sibling donor (Karlson *et al.*, 2012).

In Equatorial Africa, the prevalence of the sickle cell trait stands at 10-40%, going down to even lower rates in Northern Africa, and especially the Southern part of Africa where prevalence rates are below 1%. In West African countries such as Ghana and Nigeria, rates of about 15-30% have been recorded for children born with the sickle cell trait. SCD births are based on the number of carriers within a population, and the numbers are purported to be due to the survivor effect of carriers against malaria transmission, and hence ability to continually pass on the Sickler gene to offspring and generations (Anie *et al.*, 2010).

2.2 Clinical Variability of SCD

Sickle Cell Disease shows clinical variability, with some people showing severe forms of the disease characterized by concomitant complications as well as frequent hospitalizations, while others show no signs at all, or experience benign signs of the disease. Both genetic and environmental factors affect SCD variability, however socioeconomic status has been shown to play a major role affecting clinical manifestation, by means of feeding, practice of safe hygienic practices, as well as access to quality health care (Faremi, Olatubi, & Lawal, 2018).

Pain episodes in SCD are the main characteristic of the disease, however other clinical syndromes such as anaemia, infection, chest complications, leg ulcerations and even organ failure are recorded as effects of the disease (Ola *et al.*, 2016).

There is no known cure for SCD, and the disease is mainly managed using biomedical treatment, by the use of analgesics to reduce the pain (Ola *et al.*, 2016), as well as increase in

fluid intake, taking folic acid to supplement the body's immune responses, and limiting the amount of physical activity the individuals undertake (Karlson *et al.*, 2012). In some cases, regular blood transfusions for the individuals have been noted (Smith, 2016). SCD is thus characterized by chronic pain, disability, and several disease management strategies.

Countries in the Sub Saharan Africa region recorded several limitations in the curb of the disease with its introduction and discovery. Unavailability of access to standardized health care for sickle cell disease patients, as well as lack of new-born hemoglobinopathy screening are some of the causes of the estimated 50-90% of deaths of children with SCD (Kwarteng-Siaw *et al.*, 2017).

Countries that have well-resourced health centres, and have implemented new-born screening, as well as other interventions in the past have seen massive improvements of up to 95% in the survival of affected children from childhood to adulthood (Kwarteng-Siaw *et al.*, 2017).

2.3 Interventions Addressing SCD

In Ghana, Accra and Kumasi rolled out new-born screening programs in 2010, after pilot projects begun in 1995 showed a reduced under-five SCD mortality rate for children who had been signed onto the clinic (Dennis-antwi *et al.*, 2011).

The life expectancy and survival of persons with SCD are affected by varying factors including physiological, environmental, and psychological and social influences. Medical advancements such as biomedical treatments involving the use of hydroxoa, penicillin injections, and stem cell transplants have been of immense benefits to such individuals (Brewer, 2011).

The lifelong treatment and care for the disease, and all the manifestations of the illness can be particularly tasking on children. Despite the treatment regimen, children still require routine

hospital visits, and the different psychological, emotional and other complications also affect the general well-being of the child (Brewer, 2011).

Children who are diagnosed early at birth and who receive penicillin prophylaxis, and other vaccines, tend to have better health than those diagnosed later. Being taken care of at special sickle cell centres, and by specialist doctors, as well has a positive effect on the health of people with SCD than those treated in smaller, unspecialised centres (Dennis-Antwi, Dyson, & Ohene-Frempong, 2008). Adolescents and adults who receive good guidance from guardians early in their lives, comply with doctors' instructions concerning their health regimen, and the chronic nature of the disease fare much better than others who do not (Inati-Khoriaty, 2008).

2.4 Negative Effects of SCD

Children with SCD generally record poor QOL scores due to the interplay of several factors including financial capabilities of the families (Panepinto, Pajewski, Foerster, Sabnis, & Hoffmann, 2009), which can lead to increased severity of the disease (Pereira, Brener, Cardoso, & Proietti, 2013). Challenges at this stage of transition such as anxiety may be further exacerbated by the more traditional practice of transference of care from paediatrics to adult care, during mid-teen years (Kwarteng-Siaw *et al.*, 2017).

According to Brewer (2011), adolescents with SCD experience more limitations at this stage of transition from childhood to adulthood, as they must begin to independently care for themselves, from previous care by other people. He explains further that this involves circumventing ethnic and cultural factors, sociodemographic characteristics, factors related to the diseases, and any other comorbidities, all of which affect QOL. Again, recurring pain may also increase as children get older. The challenges experienced at this stage affect QOL, and

may have more far-reaching effects such as poor academic performance, and affect interpersonal relations as well (Brewer, 2011).

Other Disease Effects

SCD is a disease that has some emotional and physical factors that may have negative effects that may be stressing on patients as well as their families with regards to their quality of life. SCD patients are also likely to experience poor adaptation to life, in areas such as psychosocial relations, limited physical functioning, negative self- image, and problems in behaviour (Adzika, Glozah, & Ahorlu, 2016).

2.5 Adolescence Period and Relating Behaviours

Adolescence is an uncertain timeframe in any individual's life where risky behaviours that can negatively affect their health may start. This may affect them both at the time, and later on in their lives, with some of these behaviour modifications being difficult to correct. Evidence has shown that this development stage is marked by some risky health behaviours, especially the relationships between substance abuse, and sexual behaviour (Jackson, Sweeting, & Haw, 2012).

The adolescence period is marked by several stages of development. The period from 12 years to 14 years are the early adolescence years, where most teenagers undergo many developmental changes including physical, social, emotional and mental changes. As their bodies especially undergo major changes, they begin to worry about their outlook among their peers. Peer pressure also begins to set in at this stage, involving the introduction to the use of drugs, alcohol, and sex. This period has been noted for the onset of depression, and poor eating habits and disorders. They begin to make their own choices concerning major aspects of their lives,

such as school, their friendships, and start developing their own personalities, and interests (Health Reference Series, 2010).

With the many changes at this point in their lives, come its accompanying emotional challenges such as lack of confidence, high expectations, peer influence, and mental challenges such as more complex thought processes, and stronger sense of morality (Health Reference Series, 2010). Adult supervision is necessary to encourage the early adolescent to adequately navigate these challenges with the best possible outcome.

From 15-17 years old, the adolescent goes through middle adolescence, where mental, cognitive, and sexual changes are more pronounced. Teenagers become more responsible, and independent both from their parents, and their peers as well, as they begin to form a clearer sense of identity showing more concern for their future goals and vocations. It is noteworthy that the brain development at this timeframe sets the stage for priority setting, planning, concrete decision making, and impulse control, (Health Reference Series, 2010) components of taking care of one self.

2.5.1 Adolescence Stage and Health

Puberty begins at the onset of adolescence. During this timeframe of transition, males go through several changes physically, mentally, emotionally, and socially. Bodily changes including weight gain, and increase in height, as well as enlargement of sexual organs takes place. Mental changes including a sense of independence, and identity begins to be formed at this stage (Health Reference Series, 2010).

Sutton (2010) records that females tend to have more negative responses during this stage to the adjustments in their lives, than their male counterparts. The period is marked by the onset

of puberty, involving bodily changes such as the extension of breasts, and increase in height and weights, as well as more hormonal changes such as the onset of menstruation (Health Reference Series, 2010)

The adolescent period of a person's life is generally thought to be healthy. However, in developing countries, the health of adolescents is being compromised due to exposures they are not well equipped to handle (Ohene, Tettey, & Kumoji, 2011).

In discovering and developing their identities, adolescents take risks. These risks may either be healthy such as socialisation, development of their creative abilities, volunteering, taking up leadership positions. Risks taken by adolescents may also be negative such as drinking, unsafe sexual activity, smoking, and drug use. Again, adult supervision and guidance at this stage is key (Health Reference Series, 2010).

The adolescent stage is a period of development, marked by the formation of interpersonal relationships with peers, discovery of new experiences, physical development, as well as familial bonds (Forrester *et al.*, 2015).

Improving health care is not the only method to successfully adapt people to management of a chronic disease, as there are other physiological and mental challenges that have to be overcome as well. A major component of good psychological health is the building of good social and interpersonal relations from childhood into adolescence, and adapting coping mechanisms that will help prevent depression, and encourage bonding at the family level (Forrester *et al.*, 2015).

People with SCD are known to experience several other health problems associated with the disease, in terms of its psychological and physical effects. Some of these negative effects are curbed by building good social relationships from childhood to adolescence, in the advancement of their psychosocial development, an essential component of good health (Forrester *et al.*, 2015).

The challenges found at this stage of life are not unique to individuals with SCD only, as all youth have to navigate this transition period as best as they can, with all its varying life changes. Adolescents with SCD need extra support from their families and other medical personnel as they come to terms with management of the disease and committing to life with chronic illness marked by some naturally accompanying functional disabilities, for the rest of their lives (Treadwell, Telfair, Gibson, Johnson, & Osunkwo, 2017).

Relationships may be strained by the constant and protracted hospital admissions in Sickle Cell Disease, and may be the cause of the disruption of the formation of peer relationships at this stage. This may negatively affect the physical and psychological development of the adolescents at this period, as they are building bonds with their colleagues, and learning other new experiences which may be affected by complications and other effects of the disease (Forrester *et al.*, 2015).

Frequent hospitalizations, late physical development, and peer pressure at this stage of life may affect the development of the adolescent at this stage, as a result of the negative effects of the disease at such a stage. These may even lead to psychological disorders such as depression, low self-esteem and engaging in risky behaviours such as drinking, smoking, and poor sexual health practices, disorders which have been reportedly high among adolescents with SCD.

These disorders may then increase the severity of the symptoms of the disease, resulting in more, and longer hospital stays. (Forrester *et al.*, 2015).

SCD has been found to be one of the top three causes of deaths of adolescents, and was the leading cause of death among non-communicable diseases in adolescents in a study conducted by Ohene *et al.* (2011).

2.6 Sickle Cell Disease and the Adolescence Period

The last thirty years have seen SCD being recognised as a lifelong condition, as opposed to the previous perception of it being a childhood disease (Roberti *et al.*, 2010). Most children now survive into adulthood, especially in developed countries, with more advanced treatment methods. Although most young adults have been able to adapt to the adult system of care after being transferred from paediatric care, some still experiences challenges with the transition (Treadwell *et al.*, 2017).

Transition is a longitudinal process involving a beginning phase of preparation for a change. This is followed by a “transition readiness” period involving all the decisions and steps taken to build the capacity of the individual, as well as those directly linked to him/her with regards to medical care. The transition process then has an ending stage where the individual is fully grafted into the adult care system, and is actively participating in self-care. The transition period transcends health care, and affects the daily life of the individual as well (Treadwell *et al.*, 2017).

Adolescents with SCD have been found to experience problems with anxiety, and adequately accepting and adapting life to the disease, also experiencing challenges building relationships with peers, and fitting into society (Karlson *et al.*, 2012).

Adolescents with painful chronic diseases like SCD have been found to be alienated and stigmatized by various members of their immediate environment, placing a psychological burden on them. This burden may thus affect school attendance, and indirectly future employment opportunities and possibly future relationships, affecting the mental health of the individuals, and leading to more health care service access (Forrester *et al.*, 2015).

In order to be able to relate more with their peers, adolescents may take to indulging in risky behaviours, to fit in, and show some level of independence. Although adolescents spend a lot of time in the hospitals due to their routine visits, behavioural challenges such as these are hardly ever addressed (Asnani *et al.*, 2014).

Management of Sickle Cell Disease at this stage of life has been found to be more complicated, and involving many other factors, as compared to other stages of life such as early, or later life.

As they develop, adolescents tend to become more independent, and take more responsibility for themselves in taking care of themselves. This is because as they age, parents and guardians allot more responsibility to adolescents at this transition stage in handling themselves, especially with management of a chronic illness. Adolescents fall short of these expectations of their parents, and meta-analysis has shown that older adolescents demonstrated even poorer adherence to treatment regimens (Loiselle, Lee, Szulcowski, Drake, Crosby, & Pai, 2016). Due to the complexity of the stage with regards to independence, and increasing responsibility,

shared responsibility between older caretakers such as guardians, and parents and the adolescents, has been proven to be the best form of SCD care (Smith, 2016).

The effects of non-adherence to disease management strategies are direct in cases such as more frequent hospitalizations, and increased risk of early death, and more indirect in cases by affecting more daily routines such as sleep, and frequency at school (Smith, 2016).

2.7 Quality of Life

Studies have shown that children with SCD record lower health-related quality of life scores, as compared to their counterparts, and in the adolescent years show problems with anxiety, internalizing, as well as challenges with their social relationships (Boulet, Yanni, Creary, & Olney, 2010; Karlson *et al.*, 2012).

Quality of life (QOL) is defined as a multi-dimensional construct, useful for the evaluating the effects of a chronic disease on the life of an individual (Lim, 2009) as cited in Ola *et al.*(2016). This concept broadly covers how individuals describe the ‘well-being’ of different aspects of their lives. These assessments include a person’s sentiments towards life, outlook, self-actualization, and interwork and interpersonal interactions (Theofilou, 2013).

The quality of life as perceived by an individual gives evidence of how the individual is able to cope with the effects of a chronic disease such as SCD in daily life occurrences, and would thus give an indication of successes or failures in that regard.

Studies have shown that people who have long-term conditions are more likely to experience mental health challenges than their healthy counterparts (Adzika *et al.*, 2016; Lewis &

Vitulano, 2003). In Jamaica and in America, research conducted showed that patients with SCD showed higher prevalence rates for depression, than the healthier controls, with the African Americans especially showing much higher odds of suffering from depression among those with more severe clinical SCD (Asnani, Fraser, Lewis, & Reid, 2010). Again, patients with very severe and extremely high anxiety levels report more painful crises periods (Adzika *et al.*, 2016; Mahdi, Al-Ola, Khalek, & Almawi, 2010).

Determining QOL is important as it assesses treatment programs, evaluates factors for prognosis, compares therapeutic regimens, and resource allocation periods (Adzika *et al.*, 2016). Dampier, Lebeau, Rhee, & Lieff (2013) conducted a study showing that adults with SCD had impaired QOL scores. Blood transfusions, vaso-occlusive pain crises, and other SCD morbidities affected QOL of patients in a study conducted by Pereira, Brener, Cardoso, & Proietti (2013).

2.8 Quality of Life of Adolescents with SCD

This section presents analysis of the existing literature on the factors that could associate with the quality of life of SCD clients. Some of these factors have been presented below.

According to World Health Organisation (WHO), QOL involves an individual's perception of their standing in life in the context of their culture, and values, considering their life goals and expectations. This outlines the different constructs used in measuring QOL. There are 8 domains of functional health and well-being that are assessed namely; physical function, role function, bodily pain, general health, vitality, social functioning, emotional well-being and mental health, as well as other health aspects (Theofilou, 2013). Theofilou (2013) asserts that these constructs are well known, and has been validated for use of measuring quality of life for

general well-being and more specific diseased populations, allowing measurement of disease burdens, and efficacy of various treatments.

Different cultures have special ways of describing the illness experience, influencing how the illnesses are seen, how they are experienced, and valued, as well as coping mechanisms that exist to deal with the disease. Cultures have the tendency to influence health-decision making as well (Opare-henaku & Utsey, 2017). Culture may thus have some relation with quality of life. Understanding culture in this context may prove valuable in appreciating the effect of culture on adolescents with SCD (Lim, 2009).

(Thomas & Taylor, 2002) explored the experiences of people with sickle cell disease (SCD) in order to understand the psychosocial impact of the disease. These researchers concluded that ‘SCD carries a huge psychosocial burden impacting on physical, psychological, social and occupational well-being as well as levels of independence and environment’ of the client. These factors could have an effect of the QOL of the adolescent.

The stage of adolescence is experienced by both males and females differently with regards to their adjustments as assessed by (Health Reference Series, 2010). Coping additionally with disease management of a chronic ailment like SCD, as well as all the social and cultural factors may result in a difference in the perception of both male and female adolescents towards their quality of life.

2.9 Conceptual Framework of the Quality of Life of SCD Adolescents

This section presents the conceptual framework of the study after an extensive analysis of literature on the key concepts and theory as shown in figure 1.0.

2.9.1 Narrative to Support Framework

More resources are dedicated to the disease management from a biological aspect in clinical care, and not enough to the behavioural aspect of the patients involved (Asnani *et al.*, 2014).

There are several factors that come into play with regards to the quality of life at the adolescent stage with the disease.

The psychosocial difficulties listed may affect quality of life of adolescents. Research has shown that stigma and depression are associated with persons living with SCD (Ola *et al.*, 2016). Most of these arise from poor understanding of the condition by society, and peers. Discrimination, disability, due to retarded growth, an effect of SCD, as well as poor familial bonds and social relations coupled with management of a chronic illness at the adolescent age, may result in a poor quality of life for the adolescent suffering from SCD at opposed to his/her peers at this stage.

Other factors such as the disease itself, chronic joint pains, and frequent hospitalizations may also result in poor physical, social, and emotional outcomes, affecting general health and bodily pain negatively (Forrester *et al.*, 2015).

Again, the adolescent stage in itself presents varying physical, emotional, social and psychological challenges to the individual, as they transition from childhood to adulthood (Health Reference Series, 2010).

As shown by the arrow, poor quality of life may in turn lead to the same psychosocial difficulties that seemingly caused them, such as depression, and stigmatisation due to the negative responses recorded. Social relationships may be adversely affected by turning to smoking, and other deviant behaviours, and engagement in such behaviours may take its toll on finances, as some of these are expensive habits.

The family plays a role in the quality of life of the individual, in terms of shared responsibility of support, and income to support good health of the adolescent. Again, economic factors, which may affect quality of life indirectly, by the health facility factors such as quality of care,

and the lifestyle factors which include diet, exercise, and hygiene, may also affect QOL directly.

The socio-demographic characteristics of the individual, such as the age, sex, level of education, ethnicity, and religious affiliation of the adolescent are some of the variables under study, and seek to explore the extent to which quality of life is affected by these, and other factors. The framework was developed in this context considering the domains of the SF-36, a commonly used tool for determining QOL (Theofilou, 2013).

2.9 Conceptual Framework Showing factors affecting Quality of Life of Adolescents with SCD

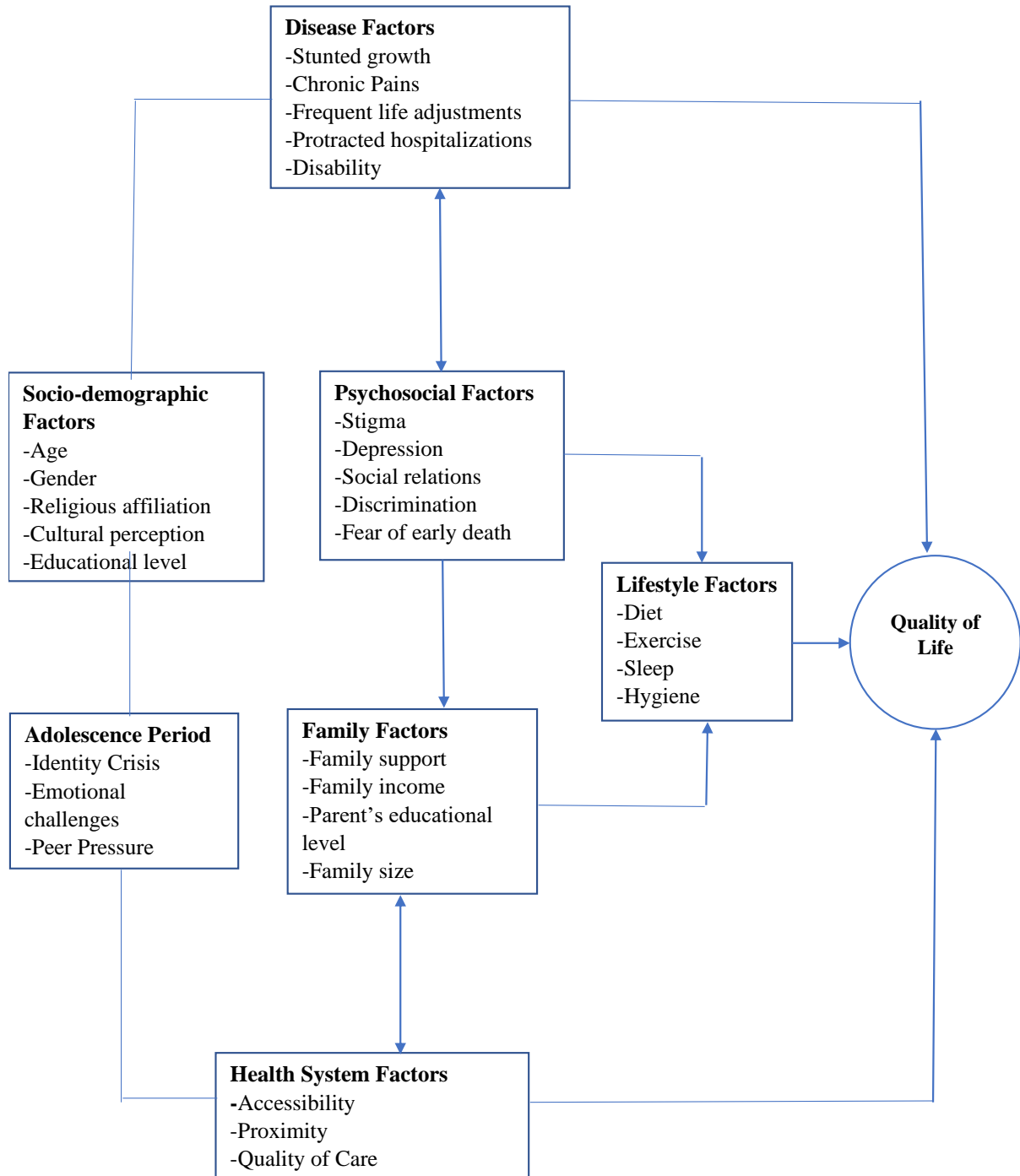


Fig. 1.0: Conceptual Framework Showing factors affecting Quality of Life of Adolescents with SCD

Author's own construct

CHAPTER THREE

METHODS

3.0 Introduction

This section details the methods and techniques that were used in conducting this study. There are nine sections. Section one describes the philosophical position with regards to the study, while section two details the study design. Section three presents the study area. Section four presents the study population, including the inclusion and exclusion criteria for the study. Section five presents the sampling technique, while section six details the data collection and seven, the analysis techniques. In section eight, the quality assurance of the study is presented, and section nine discusses the ethical considerations of the study.

3.1 Philosophical Position

The study is a qualitative research, and thus assumed the interpretivist's epistemological, and constructivist's ontological perspective. These perspectives hold that research is based on meanings people assign to data, and the subjective experiences of the world. The nature of inquiry in this paradigm was to understand a particular phenomenon, and thus quality of life in this research sought to be understood from the viewpoint of the adolescent sufferers themselves (Antwi & Kasim, 2016).

3.2 Study Design

A qualitative approach was used in this study using the phenomenological perspective. The qualitative study design is an exploratory approach that seeks to gather information on people's sense of the world, and how they ascribe meaning to their experiences (Merriam, 2009).

Qualitative research enables the researcher capture information from the data obtained in the form of the thoughts, feelings, behaviour and insights of the participants (Mayan, 2001).

Phenomenological research, deals with describing the lived experiences of individuals about a phenomenon and it culminates in the experience of multiple individuals experiencing the same phenomenon (Creswell, 2009). It allows participants to share their perceptions, feelings and lived experiences and how these affect their views concerning a given situation (Adongo *et al.*, 2016).

This approach was used in order to be able to get an in-depth view of the adolescents with Sickle Cell Disease on their perceptions concerning their physical, emotional, mental health, social functioning, and general well-being.

3.3 Study Area

Study Site:

Participants for this study were recruited at the Ghana Institute of Clinical Genetics of Korle-Bu Teaching Hospital (KBTH). The hospital is located in Accra, the capital city, in the Ablekuma South sub-metropolitan area. The Ablekuma South Sub Metropolitan is the largest in the Metropolis and shares its boundaries with Ablekuma Central, Ablekuma North and Ashiedu Keteke. It covers an area of 15.1 sqkm.

The Sub Metro has an estimated population of 257,543 with 22,751 houses and 69,401 households according the 2010 housing Census. In 2018, using the Greater Accra Growth Rate of 3.1%, it is projected that the population of Ablekuma South stands at 315,051. The sub metro has 5 electoral areas, which are; Korle Gonno, Korle Bu, Chorkor, New Mamprobi, and Mamprobi (Accra Metropolitan Assembly, 2019).

The Korle-Bu Teaching hospital is the largest tertiary referral hospital in Ghana with a current bed capacity of about 2,000. It has an average daily attendance of about 1,500 people, and

about 250 patient admissions (Korle-Bu Teaching Hospital, 2012). The Hospital also provides sophisticated and scientific investigative procedures and specialisation in various fields such as Neuro-surgery, Dentistry, Eye, ENT, Renal, Orthopaedics, Oncology, Dermatology, Cardiothoracic, Radiotherapy, Radio diagnosis, Paediatric Surgery and Reconstructive Plastic Surgery and Burns.

The study was conducted at the Ghana Institute of Clinical Genetics (Sickle Cell Clinic) located at the premises of Korle Bu Teaching Hospital. The Institute was established in 1974, and provides SCD education in the community, research in the field, as well as care to adolescents and adults with SCD. The GICG renders outpatient and emergency healthcare services to patients who report to the institute. It has a patient population of about 25,000. Yearly attendance averages about 10,000-15,000 patients, and daily attendance records are set at about 50 patients a day. The institute receives referrals from other healthcare facilities all over the country (Sey *et al.*, 2018).

3.4 Study Population

The study population consists of sickle cell adolescents aged 13-19 years reporting to the GICG without sickle cell crises. The total adolescent population of the Institute stands at about 500 patients, out of a total population of about 25,000, with average monthly adolescent attendance at about 80 patients.

3.4.1 Inclusion Criteria

The study was conducted with adolescents with SCD aged 13-18 years attending Sickle Cell clinic at GICG in July, 2019. The different genotypes present at the time of research were included in the study. Informed consent was obtained from the adults, and assent from the minors with permission and signed consent slips from their parents.

3.4.2 Exclusion Criteria

Patients aged 13-19 years who presented at the institute with any pain crises were not included in the study.

3.5 Sampling

Sampling was done among the adolescent patients who presented at the hospital for clinical check-up. The GICG provides both emergency and outpatient care for all Sickle patients referred there aged 13 years and above.

Sampling was done until data saturation was achieved, where no new information was obtained. For this research, 15 adolescents voluntarily took part in the research, with consent from their parents where necessary.

Purposive sampling technique was used to obtain participants of the study. For each day of reporting at the hospital during research, participants consisted of patients attending clinic, without crisis or acute symptoms.

3.5.1 Sampling Technique

The study was a qualitative study using the phenomenological approach. Patients who presented at the hospital without crises were considered for the study. The researcher was assisted by health staff in identifying patients who met the inclusion criteria. This technique was used until data saturation was reached involving no new information providing a sample size of 15 participants in all.

Purposive sampling was used to approach potential participants of the study informing them of the purpose of the study. They were assured of the possibility of voluntary withdrawal at any point of the study without any consequences.

3.6 Data Collection

Data was collected in July, 2019. In-depth interviews were organised for participants who agree to take part in the study. Data was collected using a semi-structured interview guide. The guide was developed in English but was translated into Twi, where this was the language the participant was comfortable with. Questions on demographics and clinical attendance were also included in the interview.

Permission was sought from the participants to audiotape the interview.

Field notes were made during the interviews, and transformed into a data document.

Data gathered was stored on the personal computer of the principal investigator. Audio recordings were deleted immediately after they had been transcribed.

The interview was conducted by the principal researcher only.

Quality of life is an index that can be measured by self-report based on their perceptions of each of the domains being measured. (Lins & Carvalho, 2016). The domains assessed in this study included general health, mental, emotional, physical, social, and cultural health and well-being, influenced by the domains assessed in the SF-36 questionnaire. Questions in the developed interview guide were based on obtaining information on these domains from study participants.

3.7 Data Analysis

The taped interviews were transcribed before analysis was done.

Thematic analysis was used employing both deductive and inductive analysis (Creswell, 2009).

A codebook containing the various codes that were used was generated based on the objectives of the study and the subject areas explored during the interviews. Major and sub-themes emerged during thematic content analysis, and a table of themes was drawn up in Microsoft word for further interpretation, and easy collation of the data. Codes were typed in Word format

for easy reading and selection of the best quotes which have been presented in the results section of the work. The major themes were then arranged based on the research questions guiding the study.

3.8 Quality Assurance

3.8.1 Trustworthiness (Rigour)

Rigour refers to the extent to which the researcher strives for excellence and how they adhere to detail and accuracy. The research used the framework by Guba and Lincoln (1985) to ensure the trustworthiness of this study. This framework encompasses the following four criteria for developing trustworthiness of a qualitative study, credibility, dependability, confirmability and transferability (Lincoln & Guba, 1985).

3.9 Ethical Considerations

Ethical issues in the conduct of a study involving patients were adhered to. These include; confidentiality, anonymity, and privacy. Participants were assured that all their information would be kept highly confidential and treated with utmost respect. Their identities were to be kept secret, and unknown only to the Principal Investigator, and identified by generically generated ID numbers. They were assured of utmost privacy in keeping their information safe.

3.9.1 Ethical Clearance

Ethical approval was sought from the Ethical Review Committee of the College of Health Sciences, supervisory body of the hospital in charge of authorising the approval of research work to be conducted for students in the college, before the start of data collection. Approval from The Ghana Institute of Clinical Genetics was also obtained before data collection began.

CHAPTER FOUR

FINDINGS

4.0 Introduction

This chapter provides the results of the research conducted on the quality of life of adolescents with Sickle Cell Disease attending the Ghana Instituted of Clinical Genetics, Korle Bu Teaching Hospital. The main objective of the study was to assess the quality of life of adolescents with SCD, to examine their perceptions of how their QOL is affected by the presence of SCD, and any socio-cultural factors of the disease, as well as to determine if there are any differences in the QOL of male and female adolescents as perceived by them. There were three main themes that emerged, with several sub-themes under each theme. The themes are presented based on the objectives the study sought to achieve.

4.1 Socio-Demographic Characteristics of Participants

Fifteen participants took part in the study with ages ranging from 15-19 years. The mean age of participants was 17.5. Six of the participants were 19 years old, while 5 of them were 17 years. 10 out of the 15 study participants were SS genotype, 3 were SC, and there was 1 CC and 1 Sickle Beta genotype participant. Average number of crisis per year was 3. There was one Muslim participant, all others were Christian. All participants were single, and literate. The tribal affiliations of the participants were required to determine cultural influences on the disease. Languages spoken were sought to ascertain ability of participants to adequately express themselves in obtaining treatment, and depth of tribal affiliation.

Table 1.0 shows the socio-demographic characteristics of participants.

ID	Age	Sex	Genotype	Average Number of Crisis per Year	Level of Education	Tribe	Languages Spoken
1	17	Male	SS	2 crisis per year	SHS	Ga	English, Ga, Twi
2	17	Male	SC	2 crisis per year	SHS	Asante	English, Ga, Twi, Hausa
3	18	Female	SC	One crisis ever	Tertiary	Gonja	English, Hausa, Twi
4	17	Male	SS	3 crisis per year	SHS	Ga	Ga, English, Twi
5	19	Female	SS	Uncountable number of crisis per year; sometimes every week.	SHS	Akuapem	English, Twi, Guan
6	19	Male	SS	2 crisis per year	SHS	Asante	English, Twi, Ga
7	19	Female	SS	3 crisis per year	SHS	Ga	English, Ga
8	16	Male	CC	5 or 6 times a year, sometimes monthly.	SHS	Ga	English, Ga, Twi
9	19	Male	SS	4 or more crisis in a year	SHS	Ewe	English, Twi.
10	19	Female	SS	Hardly ever get crisis	SHS	Ga Adangbe	English, Ga
11	17	Female	SC	Hardly ever	SHS	Ewe	English, Twi, Ga
12	15	Female	SS	Monthly crisis	JHS	Fante	English, Twi
13	15	Female	Sickle Beta	Cannot recall	SHS	Fante	English, Twi, Fante
14	17	Male	SS	2-4 crisis a year	SHS	Ga	English, Ga, a little twi
15	19	Female	SS	Hardly ever has crisis	Tertiary	Ga	English, Ga

Table 1.0: Table showing Socio-demographic Characteristics of Participants

4.2 Research Question One: What are the perceptions of adolescents with SCD on how the disease affects their quality of life?

4.2.1 Quality of Life

This objective sought to determine how the quality of life of adolescents is affected by the presence of SCD as perceived by the adolescents themselves. The quality of life concept was assessed on the basis of four main domains, namely; general health, physical health, emotional health, social and academic life, as well as their health in relation to their peers. The overall response of this finding shows that quality of life as perceived by the adolescents themselves, is good.

General Health:

Almost all of the adolescents reported their health generally as being okay, or normal. This response remained constant with participants, whether they were on the lower extremity of the spectrum of the average number of crisis, or on the higher end, reporting to the GICG almost every month for crisis pain. When probed as to what normal meant, one participant reported that;

“For me, my health is normal. Not very good, not very bad. Just okay.” (female, SS, SHS)

Another participant said; *“My health is okay. It’s not good, it’s not bad, just okay.” (female, SS, JHS)*

Physical Health:

The male participants of the study mainly reported an inability to take part in physical sports such as football, and athletics, due to the tedious nature of activities which may exacerbate their condition. While most male adolescents indicated no participation in sports activities whatsoever, a few of them mentioned engaging in less exhausting activities such as basketball:

“I don’t play football, and I don’t do athletics, but I play basketball.” (male, SS, SHS)

“Yes, my health prevents me from taking part in physical activities, like, I don’t swim. I can’t learn how to swim because of the disease. I don’t also play ball, football, even though I like football.” (male, SS, SHS)

Female adolescents mentioned lack of participation in sports mainly due to lack of interest in such: *“I don’t take part in sports because I don’t have interest in sports, not because of the disease.” (female, SC, SHS)*

Both sexes, however, did mention being exempt from exhaustive domestic chores such as sweeping, and scrubbing, especially at the Senior High School level:

“I’m not really a physical person, so it’s actually like a benefit. I didn’t take part in any housework in SHS.” (female, SC, SHS)

“No. I cannot take part in any physical activities. I cannot do them. I don’t sweep, I sweep, but when the crisis comes, I don’t sweep, I don’t do anything...” (male, SS, SHS)

“In SHS, I wasn’t required to do house chores, because of my condition.” (female, SS, SHS)

Emotional Health:

Emotional constructs assessed here were mainly happiness, depression, sadness, and mood swings. Most participants mentioned that they were always happy, and didn’t consider SCD as a major hindrance to their emotional health:

“Emotionally, I’m a very happy person. I don’t think of those things.” (female, SS, Tertiary)

“I’m usually happy, Sickle Cell doesn’t affect me emotionally”. (male, SC, SHS)

However, a few middle adolescents (15-17 years) mentioned that they struggled with coping with the disease, and were always questioning why this was happening to them, findings which were corroborated by some late adolescents about their earlier adolescence years as well.

Females suffered from such negative emotions more in this research:

“I’m always sad, and crying because of my condition. It doesn’t make you happy.” (15-year-old participant, SS, JHS)

“Now, not again, not anymore. I’ve grown out of it. Initially, somehow, I used to be moody a lot. A lot of moodiness.” (18-year-old adolescent, SC, Tertiary)

“I used to be very sad because of the sickle cell, always crying. I used to ask my parents why they gave birth to me, because I was always suffering. Because none of my parents and siblings has Sickle Cell, but I do. But now, I’m growing out of it.” (19-year old participant, SS, SHS)

Social and Academic Life

Some participants mentioned being affected academically by onset of crisis around exams period, but managing to keep up with school work. Socially, a few participants mentioned that they didn’t make/ have a lot of friends due to their condition, while some mentioned that their parents restricted them from participating in too many social activities for fear of triggering a crisis. Despite such responses, SCD was not noted to have any significant effect socially and/or academically in participants:

“Yes, SCD prevents me from socialising, because I get sick. Socially, it affects how I make friends.” (male, SC, SHS)

“My health used to affect my academic life. I used to come home from school when I fell sick, and stay at home because of crisis. I schooled in Eastern Region, and missed exams because of crisis.” (female, SS, SHS)

“SCD has not affected my life. I’m okay. I don’t go out mainly because my parents don’t allow me to do so.” (female, SS, SHS)

My condition does not affect any other aspect of my life, my friends know I have SCD. (male, CC, SHS)

Perceived Sickle Cell Health vs. Peer Health

Most adolescents asserted that the health of their peers was seemingly better than theirs as they fell sick less, and could take part in more activities. A few did mention that they thought their health, and that of their peers was generally the same:

“All my friends are usually stronger than I am.” (female, SS, SHS)

“Yes, my health is different from my peers. I find that what they’re able to do, I can’t really do same.” (male, SS, SHS)

“I think the health of my peers and that of mine is the same. We all fall sick sometimes, and then we get better.” (male, SS, SHS)

4.3 Research Question Two: What are the social-cultural factors related to with the quality of life of adolescents with SCD?

4.3.1 Socio-Cultural Factors

This objective sought to examine the social and cultural and factors that affect the quality of life of adolescents with SCD. The factors examined were the social structures such as family support, and health facility factors, as well as culture, and stigmatisation from society.

Family Support:

All participants mentioned that their families were extremely supportive of them, especially their mothers, providing for them, assisting them in taking care of themselves, in taking medication, and attending review clinic:

“My family is very supportive. They even force me to take care of myself, come to hospital. When I was born, doctors told my parents that I would not live beyond 6 years, and my mum gave up on me. But my dad didn’t, and anytime he heard of any medication to make me better, he would go and get it for me. They really support me, because even now I’m not working, but they give me money for my medicines, and transport to the hospital.” (female, SS, SHS)

“My family has not abandoned me at all. They support me a lot. My mother brings me to the hospital.” (male, SC, SHS)

Knowledge of Culture in relation to SCD

Ghana is a very cultural society, with most participants readily providing their tribes, with some belonging to more than one tribe per their parentage, and lineage. However, the findings of this study show that all the participants had no knowledge of their tribes and their perceptions with regards to diseases such as SCD:

“I don’t know Ewe culture, so I don’t know if they have any restrictions/taboo on SCD.” (male, SS, SHS)

“I don’t really know Fante culture, so I don’t know what they say about SCD.” (female, SC, SHS)

Stigmatisation

Generally, participants mentioned that they had not been stigmatised because they had Sickle Cell Disease, although a few did mention that they actually did not tell people for fear of being stigmatised:

“People know I have SCD, but they don’t treat me differently. No stigmatisation.” (male, SS, SHS)

“People know I have SCD. No one treats me differently because of my condition.” (female, SS, SHS)

“People don’t know I have SCD. I don’t want to tell anyone because Ghanaians have some perceptions of the disease, because of the way they think.” (male, SC, SHS)

Health Facility Factors

Since Sickle Cell Disease is chronic, and requires frequent hospital visitations due to crisis and/or clinical reviews, the factors of the health facility involving interaction with the health personnel was examined. All participants had positive reviews of the staff and clinic of GICG:

“The doctors and nurses are very nice over here.” (female, SS, SHS)

“This is my second time of coming here, the doctors and nurses are very friendly and motivating...I like them.” (male, SC, SHS)

4.4 Research Question Three: Are there any differences between the perceptions of male and female adolescents with SCD on their quality of life?

Male and female adolescents tend to develop, and behave differently, and thus, may have different coping mechanisms and disease management strategies. This objective sought to determine any such mechanisms resulting in differences in the QOL of adolescents. It was assessed in terms of their attendance to clinic, and their perceptions on differences in coping and disease management strategies.

Self- Reported Clinical Attendance and Adherence to Treatment Regimen

There was no significant difference in the attendance of both sexes to clinic, per responses from participants, as well as from records. Clinical attendance was mainly based on frequency of crisis, which would as well influence review visits. There was also no significant difference in how well male and female adolescents took care of themselves. Both sexes were adequately informed on what to do to take care of themselves, and with the help of their family, appeared to abide by such recommendations on dieting, dressing, and medication.

“To take care of myself, I have to wear longer clothes, even though I usually prefer wearing shorter clothes. I’m also supposed to take a lot of fruits, I’ve been asked to take a lot of fruits,

take in vegetables, and take my medication. I take Zincovit, normal para, Folic Acid, and Vitamin C and mutivite every day. That's what the doctors asked me to take.” (male participant)

“I take my medications, I take Folic Acid, and Zincovit. We drink water, but we avoid cold water, and bathing with cold water. Dress warm. I don't sleep with fan.” (female participant)

Perceptions of Adolescents on Sex Differences in Coping and Management of the Disease.

Some adolescents mentioned that they did not see any differences between how males and females coped with SCD. However, a few of them were of the view that, males had more severe crisis than females, but females took better care of themselves, and adhered to treatment regimens better than males:

“No difference between how males and females cope, but I think SCD is more severe in boys than in girls.” (male, SS, SHS)

“I don't know if boys can cope and manage SCD better than girls. I think it depends on the individual.” (female, SB, SHS)

“I think boys have a more severe sickle cell, and crisis than girls. The girls own is normal. I also think the girls would cope with the disease better.” (female, SS, SHS)

CHAPTER FIVE

DISCUSSION

5.0 Introduction

This chapter presents a discussion of the findings of this research, based on inductive and deductive coding, as well as other literature on the phenomenon under review. The discussion is based on the themes as well as sub-themes generated from the data set. There were three main themes that emerged from the analysis, which are quality of life, socio-cultural factors, and perception of sex differences on quality of life.

5.1 Quality of Life

Quality of life is an important construct to measure, in that it assesses the impact of disease on a particular individual, enabling the health profile of the individual to be drawn, the efficiency of new treatments, and evaluation of the needs of patients and their families (Lim, 2009). In this study, quality of life was assessed on physical health, emotional health, general health, other health (social and academic life), and a comparison of peer health with the health of the study participants, based on the conceptual framework that influenced the study. The different factors as outlined in the framework were used in developing questions that sought information on how they affected the overall quality of life of the adolescents. The factors measured were socio-demographic factors, factors of the disease itself such as frequent hospitalization, psychosocial factors such as stigmatization, and depression, and lifestyle factors involving diet, and exercise. Other aspects of the developed conceptual framework are health system factors including quality of care, family support and other family factors, and challenges presented by the period of adolescence, such as emotional challenges, and peer pressure.

Studies by (Atoui *et al*, 2015; Dyson *et al*, 2012), found that adolescents tend to be negatively affected in their school work resulting in them missing tests, which was consistent with our findings. However, the participants mentioned provision by their schools to catch up on school work that they missed out on, thus they were not greatly affected.

According to (Health Reference Series, 2010), the middle adolescence period from 15-17 years is marked by cognitive, mental and sexual changes, and females tend to have a more negative response to such changes. This further asserts the result of this study, where the middle adolescents, especially the females struggled emotionally with coping and adapting to living with SCD. One of the participants directly recorded an association between her crisis, and monthly periods, and stated categorically that, “...*the disease doesn't make me happy.*”

This research found that adolescents were limited in their physical activities, by themselves or their parents stopping them, findings from a study by (Mufti, Towell, & Cartwright, 2015), as well.

The quality of life of participants as perceived by them was found to be good. This appears to be in contrast with other studies that have found that QOL of adolescents with SCD is poor, and generally poorer than their health counterparts (Amr, Amin, & Al-Omair, 2011; Mufti *et al.*, 2015).

This may be due to such studies being quantitative, and thus not being able to obtain rich data from the study participants themselves. Studies have found that adolescents tend to downplay the effects of the disease on their daily functioning, in order to appear normal (Erskine, 2011; Atoui *et al.*, 2015). Although the study participants did accept that they had health limitations in the various domains measured that their peers did not have, it did not significantly change their world view of themselves, with some of them even viewing their well-being almost as good as that of their peers (Foster & Ellis, 2018).

5.2 Socio-Cultural Factors

All participants mentioned strong ties of family support which greatly aided them in taking care of themselves, in order to prevent crisis. This was consistent with a study by (Atoui *et al.*, 2015), who found that adolescents with SCD greatly relied on their parents, and siblings as well for support, when they are ill, and before then to prevent illness.

Studies by (Dyson *et al.*, 2012; Erskine, 2011; Musumadi, Westerdale, & Appleby, 2011), found that patients with SCD were stigmatized by their peers. However, participants in this study did not make any such reports, which is consistent with a research by (Atoui *et al.*, 2015). There were no significant reports of stigmatization, or being sidelined on the part of the adolescents in the research, in cases where their peers were aware of the condition, as well as when they were not aware. Atoui *et al.* (2015) purported that this may be due to a reluctance by study participants to share such information with the researcher.

Interaction with health professionals is a necessary consistent aspect of the life of persons with a chronic illness such SCD, due to the routine hospital visits. Mufti *et al.* (2015) found that interactions with different health personnel affected how patients responded to treatment. If the health workers were nice, the patients responded better. In this research, all participants responded that the health staff at the institute were very friendly, and motivated them to take good care of themselves. This may be a crucial factor in the overall reported QOL of the adolescents.

SCD as a disease mainly affects people of African descent (Ola *et al.*, 2016), and there may be certain cultural perceptions that influence adjustment to the disease. The illness experience is shaped by cultural factors, that affect how it is perceived, labeled, experienced, and valued by individuals (Lim, 2009). Although culture plays a major role in the illness experience (Dyson *et al.*, 2012) , this study did not find any effects of culture on SCD, with regards to how the adolescents perceive SCD, or seek treatment.

5.3 Perception of Different Sexes on Quality of Life

Coping has been found to be central to the quality of life of patients with long-term diseases (Lim, 2009). Lim (2009) found there to be significant gender differences between the QOL of male and female adolescents with SCD. Females had more pain responses than males. The researcher explained that this may be based on gender socialization, as well as different coping and management strategies. Again, Amr *et al.* (2011) found significant gender differences in the Health-related Quality of life of adolescents, with females showing lower scores, especially on the physical health domains. However, findings in this study did not report any significant differences between males and females and their perceptions of QOL, except on the emotional health domain. This may be due to the support of the family, and as well the role of females in adapting to emotional changes more negatively than males (Health Reference Series, 2010). Both genders demonstrated significant knowledge on their treatment regimen, and adherence to clinical visits as well.

CHAPTER SIX

CONCLUSIONS AND RECOMMENDATIONS

6.0 Introduction

The previous section presented a summary of the findings of this dissertation, and a discussion of these findings in the context of literature reviewed. This concluding chapter provides the conclusions, limitations, and recommendations for further studies for this dissertation.

6.1 Key Findings

Key findings from the research conducted include;

1. The QOL of the adolescents attending Sickle Cell Clinic at the GICG as perceived by them was found to be good. Emotional health in some younger adolescents, particularly female adolescents was found to be lacking.
2. Social factors such as family support and health facility factors involving interaction with health staff, were found to have an effect on QOL.
3. There was no difference in the perceptions of male and female adolescents with SCD on their QOL. This was shaped by their similarity in clinical attendance, as well as their adherence to treatment regimens and coping and management mechanisms of the disease.

6.2 Conclusions

This thesis sought to determine the quality of life of adolescents living with Sickle Cell Disease, who attend Sickle Cell Clinic at the Ghana Institute of Clinical Genetics. Using the phenomenological technique of qualitative research, 15 individuals were interviewed, using a semi-structured interview guide, and thematic analysis was done manually to analyse the results obtained. Findings showed that, the overall QOL as perceived by the adolescents themselves is good, with the exception of poor emotional health among middle adolescents.

QOL was determined based on physical health, general health, emotional health, other health, and the health of the adolescents in relation to their peers. Certain socio-cultural factors identified that influenced QOL are; family support, lack of stigmatization, and health facility factors including interaction with health staff at the GICG. Culture did not have any influence on QOL, per the findings of this study. There were no significant gender differences in their perceptions of QOL.

6.3 Limitation

This study was based on a homogenous sample of participants in one particular setting, and hence does not allow for generalization. Again, the study participants were purposively sampled, and thus may not be typical of all adolescents with SCD in Ghana.

6.4 Recommendations

Per the findings of this study, it is recommended that;

1. Counselling sessions should be added to routine clinical visits at the GICG to cater for poor emotional health, of female adolescents especially to avoid exacerbating the effects of the disease.
2. Social factors such as family support and health facility factors that have positive effect on QOL are encouraged. Hospital visitations should be encouraged with family members, and enabled by them. The staff of the GICG hospital should also be empowered to continue providing supportive care.
3. Further research should be undertaken in a different context, exploring adolescents in a culturally-influenced, or rural setting, to determine if there are any differences in the quality of life of such adolescents.

REFERENCES

- Accra Metropolitan Assembly. (2019). Sub Metros: Ablekuma South Sub-Metropolitan District Council. Retrieved from <https://ama.gov.gh/sub-metro-details.php?s=Mg==>
- Adongo, P. B., Tabong, P. T., Asampong, E., Ansong, J., Robalo, M., & Adanu, R. M. (2016). Beyond Knowledge and Awareness : Addressing Misconceptions in Ghana ' s Preparation towards an Outbreak of Ebola Virus Disease. *PLoS ONE*, *11*(2), 1–20. <https://doi.org/10.1371/journal.pone.0149627>
- Adzika, V. A., Glozah, F. N., & Ahorlu, C. S. K. (2016). Socio- Demographic Characteristics and Psychosocial Consequences of Sickle Cell Disease: The Case of Patients in a Public Hospital in Ghana. *International Journal of Medical and Health Sciences*, *10*(4), 192–200. <https://doi.org/1004212>
- Amr, M. A.-M., Amin, T. T., & Al-Omair, O. A. (2011). Health Related Quality of Life among Adolescents with Sickle Cell Disease in Saudi Arabia. *Pan African Medical Journal*, *8*(10), 1–16.
- Anie, K., Egunjobi, F., & Akinyanju, O. (2010). Psychosocial impact of Sickle Cell Disease: perspectives from a Nigerian setting. *Globalization and Health*, *6*(2), 1–6. Retrieved from <http://dx.doi.org/10.1186/1744-8603-6-2> P Mid:20170540 PMC id:28-36308
- Antwi, S. K., & Kasim, H. (2016). Qualitative and Quantitative Research Paradigms in Business Research : A Qualitative and Quantitative Research Paradigms in Business Research : A Philosophical Reflection, (February).
- Asnani, M. R., Bhatt, K., Younger, N., McFarlane, S., Francis, D., Gordon-Strachan, G., & Reid, M. E. (2014). Risky behaviours of Jamaican adolescents with sickle cell disease. *Hematology*, *19*(7), 373–379. <https://doi.org/10.1179/1607845413Y.0000000136>
- Asnani, M. R., Fraser, R., Lewis, N. A., & Reid, M. E. (2010). Depression and loneliness in Jamaicans with sickle cell disease. *BMC Psychiatry*, *10*(40), 7. <https://doi.org/10.1186/1471-244X-10-40>
- Atoui, M., Badr, L. K., Brand, T. D., Khoury, R., Shahine, R., & Abboud, M. (2015). The Daily Experiences of Adolescents in Lebanon With Sickle Cell Disease. *Journal of Pediatric Health Care*, *29*(5), 424–434. <https://doi.org/10.1016/j.pedhc.2015.01.012>
- Boulet, S. L., Yanni, E. A., Creary, M. S., & Olney, R. S. (2010). Health status and healthcare use in a national sample of children with sickle cell disease. *American Journal of Preventative Medicine*, *38*(4 Suppl), 528–535. <https://doi.org/10.1016/j.amepre.2010.01.003>.
- Brewer, C. A. (2011). *Factors that influence Health-Related Quality of Life in Adolescents with Sickle Cell Disease*. University of North Carolina, Chapel Hill.
- Creswell, J. W. (1998). Qualitative inquiry and research design: Choosing among five traditions. *Qualitative Health Research*. <https://doi.org/10.1111/1467-9299.00177>
- Creswell, J. W. (2009). Research design: Qualitative, quantitative, and mixed methods approaches. *Research Design Qualitative Quantitative and Mixed Methods Approaches*, *3rd*, 260.

- Dale, J. C., Cochran, C. J., Roy, L., Jernigan, E., & Buchanan, G. R. (2011). Health-related Quality of Life in Children and Adolescents With Sickle Cell Disease. *Journal of Pediatric Health Care*, 25(4), 208–215. <https://doi.org/10.1016/j.pedhc.2009.12.006>
- Dampier, C., Lebeau, P., Rhee, S., & Lieff, S. (2013). Health-Related Quality of Life in Adults with Sickle Cell Disease (SCD): A Report from the Comprehensive Sickle Cell Centers Clinical Trial Consortium, 86(2), 203–205. <https://doi.org/10.1002/ajh.21905>.Health-Related
- Dennis-antwi, J. A., Culley, L., Hiles, D. R., Dyson, S. M., Culley, L., Hiles, D. R., & Dyson, S. M. (2011). ‘ I can die today , I can die tomorrow ’: lay perceptions of sickle cell disease in Kumasi , Ghana at a point of transition, 7858. <https://doi.org/10.1080/13557858.2010.531249>
- Dennis-Antwi, J. A., Dyson, S., & Ohene-Frempong, K. (2008). Healthcare provision for sickle cell disease in Ghana : challenges for the African context. *Diversity in Health and Social Care*, 5, 241–254.
- Dyson, S. E., Rm, R. N., Atkin, K., Dphil, B. A., Culley, L., & Ma, B. A. (2012). School ethos and variation in health experience of young people with sickle cell disorder at school. *Diversity and Equity in Health and Care*, 9, 33–44.
- Edwin, A. K., Edwin, F., & Etwire, V. (2011). Controlling sickle cell disease in Ghana - Ethics and options. *Pan African Medical Journal*, 10, 1–9. <https://doi.org/10.4314/pamj.v10i0.72223>
- Erskine, R. (2011). Adolescent boys with sickle cell disease: A qualitative study. *Clinical Child Psychology and Psychiatry*, 17, 17–31. <https://doi.org/10.1177/1359104511399843>.
- Faremi, A. F., Olatubi, M. I., & Lawal, Y. R. (2018). Knowledge of Sickle Cell Disease and Pre Marital Genotype Screening among Students of a Tertiary Educational Institution in South Western Nigeria. *International Journal of Caring Sciences*, 11(1), 285–295. Retrieved from <http://search.ebscohost.com/login.aspx?direct=true&db=cin20&AN=129399266&site=e-host-live>
- Forrester, A. B., Barton-Gooden, A., Pitter, C., & Lindo, J. L. M. (2015). The lived experiences of adolescents with sickle cell disease in Kingston, Jamaica. *International Journal of Qualitative Studies on Health and Well-Being*, 10, 28104. <https://doi.org/http://dx.doi.org/10.3402/qhw.v10.28104>
- Foster, N., & Ellis, M. (2018). Sickle cell anaemia and the experiences of young people living with the condition. *Nursing Children and Young People*, 30(3), 36–43. <https://doi.org/10.7748/ncyp.2018.e935>
- Fuggle, P., Shand, P. A. X., Gill, L. J., & Davies, S. C. (1996). Pain, quality of life, and coping in sickle cell disease. *Archives of Disease in Childhood*, 75(4-B), 199–203. Retrieved from <http://search.ebscohost.com/login.aspx?direct=true&db=psych&AN=2010-99200-380&site=e-host-live>
- Health Reference Series. (2010). *Adolescent Health Sourcebook*. (A. Sutton, Ed.) (Third Edit). Detroit: Peter E. Ruffner.
- Inati-Khoriaty, A. (2008). *Sickle Cell Disease*. (A. Eleftheriou & M. Angastiniotis, Eds.) (Publicatio). Nocosia, Cyprus: Thalassaemia International Federation.

- Jackson, C., Sweeting, H., & Haw, S. (2012). Clustering of substance use and sexual risk behaviour in adolescence: analysis of two cohort studies, 1–10. <https://doi.org/10.1136/bmjopen-2011-000661>
- Karlson, C. W. ., Leist-Haynes, S. ., Smith, M. ., Faith, M. A. ., Elkin, T. D. . b, & Megason, G. . (2012). Examination of risk and resiliency in a pediatric sickle cell disease population using the psychosocial assessment tool 2.0. *Journal of Pediatric Psychology*, *37*(9), 1031–1040. <https://doi.org/10.1093/jpepsy/jss087>
- Korle-Bu Teaching Hospital. (2012). Annual Report of the Korle Bu Teaching Hospital. Accra, Ghana: Korle Bu Teaching Hospital.
- Kwarteng-Siaw, M., Paintsil, V., Korankye Toboh, C., Owusu-Ansah, A., & S. Green, N. (2017). Assessment of Transition Readiness in Adolescents with Sickle Cell Disease and their Caretakers, A single institution experience. *International Journal of Hematology Research*, *3*(1), 171–179. <https://doi.org/10.17554/j.issn.2409-3548.2017.03.47>
- Lewis, M., & Vitulano, L. (2003). Biopsychosocial issues and risk factors in the family when the child has a chronic illness. *Child and Adolescent Psychiatric Clinics of North America*, *12*(3), 389–399. [https://doi.org/10.1016/S1056-4993\(03\)00024-5](https://doi.org/10.1016/S1056-4993(03)00024-5)
- Lim, C. M. S. (2009). *Pain, Quality of Life, and Coping in Paediatric Sickle Cell Disease. Dissertation, Georgia State University.* Georgia State University. <https://doi.org/10.1136/adc.75.3.199>
- Lins, L., & Carvalho, F. M. (2016). SF-36 total score as a single measure of health-related quality of life: Scoping review. *SAGE Open Medicine*, *4*, 205031211667172. <https://doi.org/10.1177/2050312116671725>
- Loiselle, K., Lee, J. L., Szulczewski, L., Drake, S., Crosby, L. E., & Pai, A. L. H. (2016). Systematic and Meta-Analytic Review : Medication Adherence Among Pediatric Patients With Sickle Cell Disease. *Journal of Pediatric Psychology*, *41*(September 2015), 406–418. <https://doi.org/10.1093/jpepsy/jsv084>
- Mahdi, N., Al-Ola, K., Khalek, N., & Almawi, W. (2010). Depression, Anxiety and Stress Comorbidities in Sickle Cell Anaemia Patients with Vaso-Occlusive Crisis. *J. Pediatr Hematol Oncol*, *32*(5), 345–349.
- Mayan, M. J. (2001). An Introduction to Qualitative Methods: A Training Module for Students and Professionals. *International Institute for Qualitative Methodology*.
- Merriam, S. (2009). *Qualitative research: A guide to design and implementation.* Jossey-Bass. San Francisco, CA.
- Modell, B., & Darlison, M. (2008). Public health reviews Global epidemiology of haemoglobin disorders and derived service indicators, 036673(September 2006). <https://doi.org/10.2471/BLT.06.036673>
- Mufti, G., Towell, T., & Cartwright, T. (2015). Pakistani Children’s Experiences of Growing Up With Beta-Thalassemia Major. *Qualitative Health Research*, *25*(3), 386–396. <https://doi.org/10.1177/1049732314552663>
- Musumadi, L., Westerdale, N., & Appleby, H. (2011). An overview of the effects of sickle cell disease in adolescents. *Nursing Standard*, *26*(26), 35–40. <https://doi.org/10.7748/ns2012.02.26.26.35.c8973>

- Ohene, S., Tettey, Y., & Kumoji, R. (2011). Cause of death among Ghanaian adolescents in Accra using autopsy data. *BMC Research Notes*, *4*(1), 353. <https://doi.org/10.1186/1756-0500-4-353>
- Ola, B. A. (2016). *Living With Sickle Cell and Depression in Lagos, Nigeria*. Leicester University.
- Ola, B. A., Yates, S. J., & Dyson, S. M. (2016). Living with sickle cell disease and depression in Lagos, Nigeria: A mixed methods study. *Social Science and Medicine*, *161*, 27–36. <https://doi.org/10.1016/j.socscimed.2016.05.029>
- Opare-henaku, A., & Utsey, S. O. (2017). Culturally prescribed beliefs about mental illness among the Akan of Ghana. *Transcultural Psychiatry*, *54*(4), 502–522. <https://doi.org/10.1177/1363461517708120>
- Panepinto, J. A., Pajewski, N. M., Foerster, L. M., Sabnis, S., & Hoffmann, R. G. (2009). Impact of family income and sickle cell disease on the health-related quality of life of children, 5–13. <https://doi.org/10.1007/s11136-008-9412-8>
- Pereira, S. A. dos S., Brener, S., Cardoso, C. S., & Proietti, A. B. de F. C. (2013). Sickle Cell Disease: quality of life in patients with hemoglobin SS and SC disorders. *Revista Brasileira de Hematologia e Hemoterapia*, *35*(5), 325–331. <https://doi.org/10.5581/1516-8484.20130110>
- Piel, F. ., Hay, S. I., Gupta, S., Weatherall, D. J., & Williams, T. N. (2013). Global Burden of Sickle Cell Anaemia in Children under Five, 2010–2050: Modelling Based on Demographics, Excess Mortality, and Interventions. *PLoS Medicine*, *10*(7). <https://doi.org/doi:10.1371/journal.pmed.1001484>
- Roberti, M. do R., Moreira, C., Tavares, R., Filho, H., Humberto, C., & Maia, G. (2010). Evaluation of quality of life of patients with sickle cell disease in a General Hospital of Goiás , Brazil. *Revista Brasileira de Hematologia e Hemoterapia*, *32*(6), 449–454.
- Sey, F., Benneh-Akwasi Kuma, A. A., Wilson, I., Asare, E. V., Olayemi, E., & Dei-Adomakoh, Y. (2018). Burden of Sickle Cell Disease in Ghana: The Korle-Bu Experience. *Advances in Hematology*, *2018*, 1–5. <https://doi.org/10.1155/2018/6161270>
- Smith, A. W. (2016). Parent and Adolescent Factors Related to Adherence and Health Outcomes in Sickle Cell Disease. Dissertation, Kent State University.
- Theofilou, P. (2013). Quality of life: Definition and measurement. *Europe's Journal of Psychology*, *9*(1), 150–162. <https://doi.org/10.5964/ejop.v9i1.337>
- Thomas, V. J., & Taylor, L. M. (2002). The psychosocial experience of people with sickle cell disease and its impact on quality of life: Qualitative findings from focus groups. *British Journal of Health Psychology*, *7*(3), 345–363. Retrieved from <https://doi.org/10.1348/135910702760213724>.
- Treadwell, M., Telfair, J., Gibson, R. W., Johnson, S., & Osunkwo, I. (2017). Transition from pediatric to adult care in sickle cell disease: Establishing evidence-based practice and directions for research. *American Journal of Hematology*, *86*(1), 116–120. <https://doi.org/10.1002/ajh.21880>.
- World Health Organization (WHO). (2006). *Sickle-cell anaemia Report by the Secretariat*. World Health Organization (Vol. A59). Fifty-Ninth World Health Assembly.

APPENDICES

SCHOOL OF PUBLIC HEALTH COLLEGE OF HEALTH SCIENCES UNIVERSITY OF GHANA

Appendix A: Participant's Information Sheet

(A copy of this form was given to participants aged 18 years and over)

Title of Dissertation: Quality of life of adolescents with Sickle Cell Disease attending the Ghana Institute of Clinical Genetics (Korle Bu Teaching Hospital).

Background of the study

My name is Mary Fynn, a Master of Public Health student of University of Ghana School of Public Health. This study is being conducted on the topic: **“Quality of life of adolescents with Sickle Cell Disease attending the Ghana Institute of Clinical Genetics” (Korle Bu Teaching Hospital)**. The purpose of this study is to measure the quality of life of adolescents with Sickle Cell Disease who attend clinic at the GICG. This informed consent is to ensure that you understand the purpose and your responsibilities in the research before you decide whether to participate or not. This study is part of the requirements for the award of master of public health degree. I would like to seek your approval and permission to ask some questions on the quality of life of adolescents.

Procedures/nature

Participants will fill out short questionnaires based on constructs describing quality of life.

Risks and benefits of the study

The study will not pose any harm to participants as there will be no invasive procedure. Participants will not be given any money for participation in this research whiles benefits that may arise will be a greater contribution to Sickle cell adolescent care.

The information will also arouse the interest of policy makers to pay more attention to how adolescents with SCD are cared for, and channel intervention where necessary. The study will not be of any risk or cost to you because the cost has been taken care of and your identity protected as well.

Voluntary participation/withdrawal

Participation is voluntary and you can at any point choose to withdraw from the study.

Anonymity and Confidentiality

You are assured that all information provided during this research will be taken with strict confidentiality, protected as much as possible and will be purely for research purpose. No discussion will be held regarding the research outside the team. All information will be treated as confidential as names will not be mentioned. Data collected and storage will be done by Mary Fynn, the principal investigator.

Mary Fynn

School of Public Health,

College of Health Sciences,

University of Ghana,

Post Office Box LG13,

Legon – Accra.

0271860480

Appendix B: Participant's Consent Form

Consent Form

Please initial each of the following statements to indicate your agreement:

---I agree to participate in this research study

---My participation in the research is of my own free will

---I have been informed about the purposes of the study and what participation involves

---I have had the opportunity to ask any questions I wish

---I understand that I can withdraw from the study at any time, without giving a reason and with no adverse consequences

Participant signature:

Date:

Researcher signature:

Date:

Appendix C: Parental Consent Form

Introduction

(A copy of this form was given to parents of potential participants who are minors)

Title of Dissertation: Quality of life of adolescents with Sickle Cell Disease attending of the Ghana Institute of Clinical Genetics (Korle Bu Teaching Hospital)

Background of the study

My name is Mary Fynn, a Master of Public Health student of University of Ghana School of Public Health. This study is being conducted on the topic: **“Quality of life of adolescents with Sickle Cell Disease attending the Ghana Institute of Clinical Genetics” (Korle Bu Teaching Hospital)**. The purpose of this study is to measure the quality of life of adolescents with Sickle Cell Disease who attend clinic at the GICG. This informed consent is to ensure that you understand the purpose and your responsibilities in the research before you decide whether to participate or not.

Your child has been invited to join a research study to look at “Quality of Life of Adolescents with Sickle Cell Disease: A Case Study of the Institute of Clinical Genetics.” Please take whatever time you need to discuss the study with your family and friends, or anyone else you wish to. The decision to let your child join, or not to join, is up to you.

This study is part of the requirements for the award of master of public health degree. I would like to seek your approval and permission to ask your child some questions on the quality of life of adolescents.

Procedures/nature

Your child will answer some questions based on constructs describing quality of life. This will take about 25-30 minutes.

Risks and benefits of the study

The study will not pose any harm to your child as there will be no invasive procedure. Participants will not be given any money for participation in this research whiles benefits that may arise will be a greater contribution to Sickle cell adolescent care.

The information will also arouse the interest of policy makers to pay more attention to how adolescents with SCD are cared for, and channel intervention where necessary. The study will not be of any risk or cost to you because the cost has been taken care of and your child's identity protected as well.

Voluntary participation/withdrawal

Participation is voluntary and you can choose at any point choose to withdraw your child from the study.

Anonymity and Confidentiality

You are assured that all information provided during this research will be taken with strict confidentiality, protected as much as possible and will be purely for research purpose. No discussion will be held regarding the research outside the team. All information will be treated as confidential as names will not be mentioned. Data collected and storage will be done by Mary Fynn, the principal investigator.

Mary Fynn

School of Public Health,

College of Health Sciences,

University of Ghana,

Post Office Box LG13,

Legon – Accra.

Tel. No: 0271860480

PERMISSION FOR A CHILD TO PARTICIPATE IN RESEARCH

As parent or legal guardian, I authorize _____ (child's name) to become a participant in the research study described in this form.

Child's Date of Birth

Parent or Legal Guardian's Signature

Date

(Upon signing, the parent or legal guardian will receive a copy of this form, and the original will be held in the subject's research record.)

Appendix D: Participant's Assent Form

(For Minors)

Introduction

My name is Mary Fynn and I am from the School of Public Health at the University of Ghana. I am conducting a research study entitled, "Quality of Life of Adolescents with Sickle Cell Disease attending the Ghana Institute of Clinical Genetics." I am asking you to take part in this research study because I am trying to learn more about quality of life of adolescents attending clinic at this institute. This will take about 25-30 minutes.

General Information

If you agree to be in this study, you will be asked to participate in an interview.

Possible Benefits

Your participation in this study will result in added knowledge on adolescent patient care at the GICG.

Possible Risks and Discomforts

However, the risks associated are some emotional discomfort.

Voluntary Participation and Right to Leave the Research

You can stop participating at any time if you feel uncomfortable. No one will be angry with you if you do not want to participate.

Confidentiality

Your information will be kept confidential. No one will be able to know how you responded to the questions and your information will be anonymous.

Contacts for Additional Information

You may ask me any questions about this study. You can call me at any time 0271860480 or talk to me the next time you see me.

Please talk about this study with your parents before you decide whether or not to participate.

I will also ask permission from your parents before you are enrolled into the study. Even if your parents say “yes” you can still decide not to participate.

VOLUNTARY AGREEMENT

By making a mark or thumb printing below, it means that you understand and know the issues concerning this research study. If you do not want to participate in this study, please do not sign this assent form. You and your parents will be given a copy of this form after you have signed it.

This assent form which describes the benefits, risks and procedures for the research titled “Quality of Life of Adolescents with Sickle Cell Disease: A Case Study of the Ghana Institute of Clinical Genetics” has been read and or explained to me. I have been given an opportunity to have any questions about the research answered to my satisfaction. I agree to participate.

Child’s Name:

Child’s Mark/Thumbprint:

Date:

Researcher’s Name:

Researcher’s Signature:

Date:

Appendix E: Interview Guide

SOCIO-DEMOGRAPHIC CHARACTERISTICS

- 1) How old are you? (Age at last birthday)
- 2) Sex
- 3) Genotype/Hb Status....
- 4) Average number of crisis per year.....
- 5) What is your religious affiliation?.....
- 6) What is your marital status?.....
- 7) What is your level of education?.....
- 8) Which tribe do you belong to?.....
- 9) What languages do you speak?

Main Questions

1. In general, how would you describe your health?
2. Does your health limit you in carrying out any physical activities?
3. Has your condition affected you recently in terms of your emotions?
4. How different do you think your health is from your peers?
5. Does your condition affect any other aspect of your life?
6. Do you think there are any differences between how male and females cope with adolescence and management of Sickle Cell Disease?
7. How often do you attend sickle cell clinic?
8. How do the doctors and nurses treat you?
9. What is your treatment regimen like?
10. Do you experience any stigma as a result of having SCD?
11. Do you have any restrictions/taboos because of your condition?
12. How has your family supported you in this period?



UNIVERSITY OF GHANA
COLLEGE OF HEALTH SCIENCES
ETHICAL AND PROTOCOL REVIEW COMMITTEE

June 26, 2019

Ref. No.: EPRC/JUNE/2019

Mary Ama Meyiwa Fynn
Dept. of Social and Behavioral Sciences
School of Public Health
Legon.

ETHICAL CLEARANCE

Protocol Identification Number: CHS-ET/M11 - 5.2/2018-2019

FWA: 000185779

IORG: 0005170

IRB: 00006220

The College of Health Sciences Ethical and Protocol Review Committee (EPRC) at its June, 26 2019 full board meeting reviewed and approved your re-submitted research protocol.

Title of Protocol: "Quality of Life of Adolescents with Sickle Cell Disease Attending the Ghana Institute of Clinical Genetics, at Korle Bu Teaching Hospital"

Principal Investigator: Mary Ama Meyiwa Fynn

This approval requires that you submit six-monthly review report(s) of the study to the Committee and a final full review report to the EPRC at the completion of the study. The Committee may observe, or cause to be observed, procedures and records of the study before, during and after implementation.

Please note that any significant modification(s) to this project/study must be submitted to the Committee for review and approval before its implementation.

You are required to report all serious adverse events related to this study to the EPRC within seven (7) days verbally and fourteen (14) days in writing.

As part of the review process, it is the Committee's duty to review the ethical aspects of any manuscript that may be produced from this study. You will therefore be required to furnish the Committee with any manuscript for publication.

This ethical clearance is valid till June 26, 2020.

Please always quote the protocol identification number in all future correspondence in relation to this protocol.

Signed:

Professor Andrew Anthony Adjei
Chair, Ethical and Protocol Review Committee

cc: Provost, CHS
Dean, SPH
Head, Department of Social and Behavioral Sciences