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TITLE: Patterns of illness among children with sickle cell disease accessing care at the Komfo Anokye Teaching Hospital (KATH), Kumasi, Ghana.

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KUMASI, GHANA.

PATTERNS OF ILLNESS AMONG CHILDREN WITH SICKLE CELL DISEASE ACCESSING CARE AT THE KOMFO ANOKYE TEACHING HOSPITAL (KATH), KUMASI, GHANA.

BY

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A thesis submitted to the Department of Health Promotion and Education, School of Public Health, College of Health Sciences, Kwame Nkrumah University of Science and Technology, in partial fulfilment of the requirements for the degree of Master of Public Health in Health Promotion and Education.

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DECLARATION

I hereby do declare that, except for references to other people's work which have been duly acknowledged, this piece of work is my own composition and neither in whole nor in part has this work been presented for the award of a degree in this University or elsewhere.

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DEFINITION OF TERMS

Sickle Cell Disease – A group of red blood cell disorders

Hand-foot-syndrome- Swelling of the hands and feet in infants and young children

Vaso-occlusive crisis- mild to severe pain that can start suddenly and last any length of time

Anaemia – Low levels of healthy red blood cells to carry oxygen throughout the body.

Acute Chest Syndrome – Chest pains, coughing, difficulty in breathing and fever **Splenic**

Sequestration – Spleen enlargement due to trapping of sickle cells in it.

Stroke – Sickle cells getting stuck in blood vessels and clogging blood flow to the brain

Priapism – Painful erection of the penis

Place of Residence – Where one lives over a period of time.

Age – Age in months as at last birth date

Haemoglobin Genotype – Types of haemoglobin

Sex – Biological differentiation between male and female

Haemolytic crises – Rapid destruction of large numbers of red blood cells

Malaria – Parasite causing blood disease transmitted through the bite of the anopheles mosquito

Septicaemia – Infection in which large amounts of bacteria are present in the blood

Pneumonia – Lung inflammation by bacteria or viral infection

ABREVIATIONS/ACRONYMS

SCD	Sickle Cell Disease
Hb SS	Haemoglobin genotype SS **Hb**
SC	Haemoglobin genotype SC
ACS	Acute Chest Syndrome
CDC	Centre for Disease Control and Prevention
KATH	Komfo Anokye Teaching Hospital
KNUST	Kwame Nkrumah University of Science and Technology
MOH	Ministry of Health
MCHH	Maternal and Child Health Hospital
SS	Splenic Sequestration
AOR	Adjusted Odds Ratio

ABSTRACT

Children with Sickle Cell Disease (SCD) suffer from several forms of complications. Some chronic, others acute which result in their frequent visits to the hospital. SCD is most prevalent in Sub-Saharan Africa where malaria is very common. Reports from focused studies carried out in Ghana, indicate a carrier rate ranging from 10% to 30% in the population whereas 2% of all new-borns have SCD, with the common genotypes being HbSS and HbSC. Reliable data on the common presentations and accompanying
complications presented are lacking. This study was therefore carried out to identify the patterns of Illness presented by children with SCD to the Komfo Anokye Teaching Hospital (KATH).

This was a cross-sectional study that involved the use of secondary data from the Sickle Cell Clinic at KATH for a period of three years (2012- 2014). Data from children aged ≤ 14 years with SCD accessing care at the hospital were extracted and analysed on the basis of demographic details, haemoglobin genotype, types of crises, associated infections, diagnosis and number of visits within the period. Stata software version 11 was used to test for associations between demographic factors and development of crises.

Altogether 234 children were recruited within the study period. The common crises presented were vaso-occlusive (44%) followed by haemolytic crises (35%). The most common associated infection was malaria (20.8%). The study revealed significant association of age, gender and co-morbidity to the development of SCD crises. Being in the age group
10-14, [AOR= 4.8, 95% CI, 2.16- 10.99: p-value <0.001] and having malaria as an infection, [AOR=4.7, 95% CI, (1.60- 13.86): p-value=0.005], had a statistically significant association with developing pain episode. Male gender was associated with developing haemolytic crisis [AOR=2.3, 95% CI, 1.09 – 4.91: p-value=0.027].

Sickle Cell Disease presents with several complications that may be associated with age, gender and current associated infections. Knowledge of one's state and prevention of intercurrent illnesses could result in a reduction in complications.

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CHAPTER ONE

1.0 INTRODUCTION

1.1 Background

Children with Sickle Cell Disease (SCD) suffer from several forms of complications. Some, acute while others chronic and may sometimes occur suddenly (Ebert et al, 2010) which can result in their frequent visits to the hospital (Ikefuna & Imodi, 2007) increasing the financial burden on the caretakers of these children. Sickle cell disease (SCD) is a group of disorders that affect haemoglobin, the molecule in red blood cells that transport oxygen to cells throughout the body. People with this condition have abnormal haemoglobin molecules called haemoglobin S, which can distort red blood cells into a sickle, or crescent shape.

SCD is most dominant in Sub-Saharan Africa where malaria is common. The genotypes characterized by HbSS and HbSC are dominant in the sickle cell disease population of Ghana. Reports from Ghanaian studies indicate a carrier rate in the population of 30% whereas 2% of Ghanaian new-borns have sickle cell disease (Ohene-Frimpong, 2008). According to Piel et al., (2013), new-borns with sickle cell disease is increasing globally and in Ghana, an estimated 5,815 new-borns had sickle cell disease in 2010. This figure is projected to increase to 6,855 in 2050 if no serious interventions are put in place. This will definitely increase the disease burden on the country as well as its associated complications.

Clinical manifestations of sickle cell disease usually begin in early childhood. Dactylitis (pain and/or swelling of the hands or feet) in infants and young children is often the earliest manifestation of sickle cell disease. These clinical manifestations come about when sickle blood tries to move through blood vessels and are blocked due to their sickled nature. When red blood cells sickle, they break down prematurely, which can lead to anaemia. Anaemia can cause shortness of breath, fatigue, and delayed growth and development in children. The rapid breakdown of red blood cells may also cause yellowing of the eyes and skin, which are signs

of jaundice. These episodes deprive tissues and organs of oxygen-rich blood and can lead to organ damage, especially in the lungs, kidneys, spleen, and brain. A particularly serious complication of sickle cell disease is high blood pressure in the blood vessels that supply the lungs (pulmonary hypertension). Pulmonary hypertension occurs in about onethird of adults with sickle cell disease and can lead to heart failure (Gladwin et al, 2004).

Clinical consequences can be divided into four major categories: haemolysis and haematological complications, vaso-occlusion, infections and organ dysfunction (Makani et al, 2013). However in developing countries like Ghana common morbid conditions associated with SCD are vaso-occlusive episodes, infections, Acute Chest Syndrome (ACS), and Stroke (Ansong et al, 2013). The most common clinical manifestation of SCD is the occurrence of vaso-occlusive episodes. These are characterized clinically by pain in the affected sites of the body. Pain occurrences are most often unpredictable but can be prevented and in more than 80% of the episodes, there is no potential precipitating cause (Smith and Scherer, 2010). Acute Chest Syndrome can be dangerous and should be treated in a hospital. Symptoms and signs are similar to that of pneumonia and include chest pains, cough, difficulty in breathing and fever. Strokes can also happen if sickle cells get stuck in a blood vessel and clog blood flow to the brain. About 10% of children with SCD will have a symptomatic stroke. Stroke can cause learning problems and lifelong disabilities (CDC, 2013).

Precipitating factors for some of the clinical events experienced by children with sickle cell disease may be varied with infections having been identified as a major contributor to illhealth and even death in children although the presence of the sickle cell gene on its own increases the child's risk of infections especially to certain bacterial pathogens. Traditionally, infections are a major cause of morbidity and mortality particularly in children and have been associated with the death of between 20-50% of children with sickle cell disease (Booth et al, 2010). Poor

socio- economic factors as well as poor hygienic conditions are some of the predisposing factors to these infections (Jain et al, 2013).Other factors such as dehydration, high or low temperature, high wind speed and low humidity have been identified to be precipitating factors for vaso- occlusive crisis in children with sickle cell disease (Sickle Cell Anaemia Control Project, 2012).

The chronic nature and the frequent visits of children with sickle cell disease and their caretakers to the hospitals and the poorer health status of these children affect the quality of their life and that of their parents (Van Den Tweel et al, 2008). Mothers of children with sickle cell disease have regularly reported daily emotional challenges such as being scared of losing their children, loss of control over their lives and being helpless in such situations (Burnes et al., 2008). Most often the financial burden on these families is huge and taking care of children with sickle cell disease often disrupted family interactions (Brown et al., 2010) and brought untold hardships to these families.

To reduce the burden of this condition and its complications on children, their families and the country as a whole there is the need for public health officials to plan interventional measures. There is therefore the need to know and understand the morbidity patterns of children with this disease. Data on the causes and patterns of disease complications presented to the various hospitals by children with sickle cell disease will be very relevant in determining the most common conditions that are presented, which will aid in planning preventive strategies and deciding on the amount of resources that should be allocated to the prevention of such complications (Eck et al, 2006).

1.2 Problem Statement

Sickle cell disease has recently been branded a worldwide public health problem (WHO,

2010) with recent epidemiological indications pointing to a worldwide neonatal incidence of 294,000-330,000. In Sub-Saharan Africa and in Ghana the estimated number of children under 5 with SCD in 2010 were 242,187 and 5,815 respectively and likely to increase to 352,533 and 6,855 respectively by the year 2050 (Piel et al., 2013). This situation has called for measures to be put in place to reduce its impact on the sufferers as well as the care takers of these children, not forgetting the financial burden on individuals, families', communities and the country as a whole. Interventions such as, early screening and the administration of prophylaxis, comprehensive care coordination, family-patient education and chronic red blood cells transfusion may be implemented to reduce morbidity and mortality (OheneFrimpong et al., 2008).The public health impact when these interventions are implemented will include a reduction in mortality and prolongation of life of an estimated 5,302,900 children with sickle cell disease by the year 2050. While a large scale universal screening could save the lives of up to 9,806,000 new-borns with sickle cell disease globally, 85% of whom will be born in Sub-Saharan Africa (Piel et al., 2013).

The reason for diagnosing sickle cell disease (SCD) in new-borns is to ensure that health care providers educate parents of children with SCD about the special needs of these children and also begin preventive treatment before they start developing complications. The research project entitled "New-born Screening for Sickle Cell Disease" in Ghana was launched in April 1993 and started screening in 1995. Komfo Anokye Teaching Hospital (KATH) is one of the hospitals where the project of new-borns screening for sickle cell disease was started, by 2004 a total of 9,700 sickle cell patients have been enrolled into the KATH sickle cell clinic. These patients visit the hospital once a week when they have complications. In spite of the existence of a Sickle Cell Clinic after almost 20 years of establishment of the newborn screening program, there is paucity of data published on patterns of illness among these patients. The

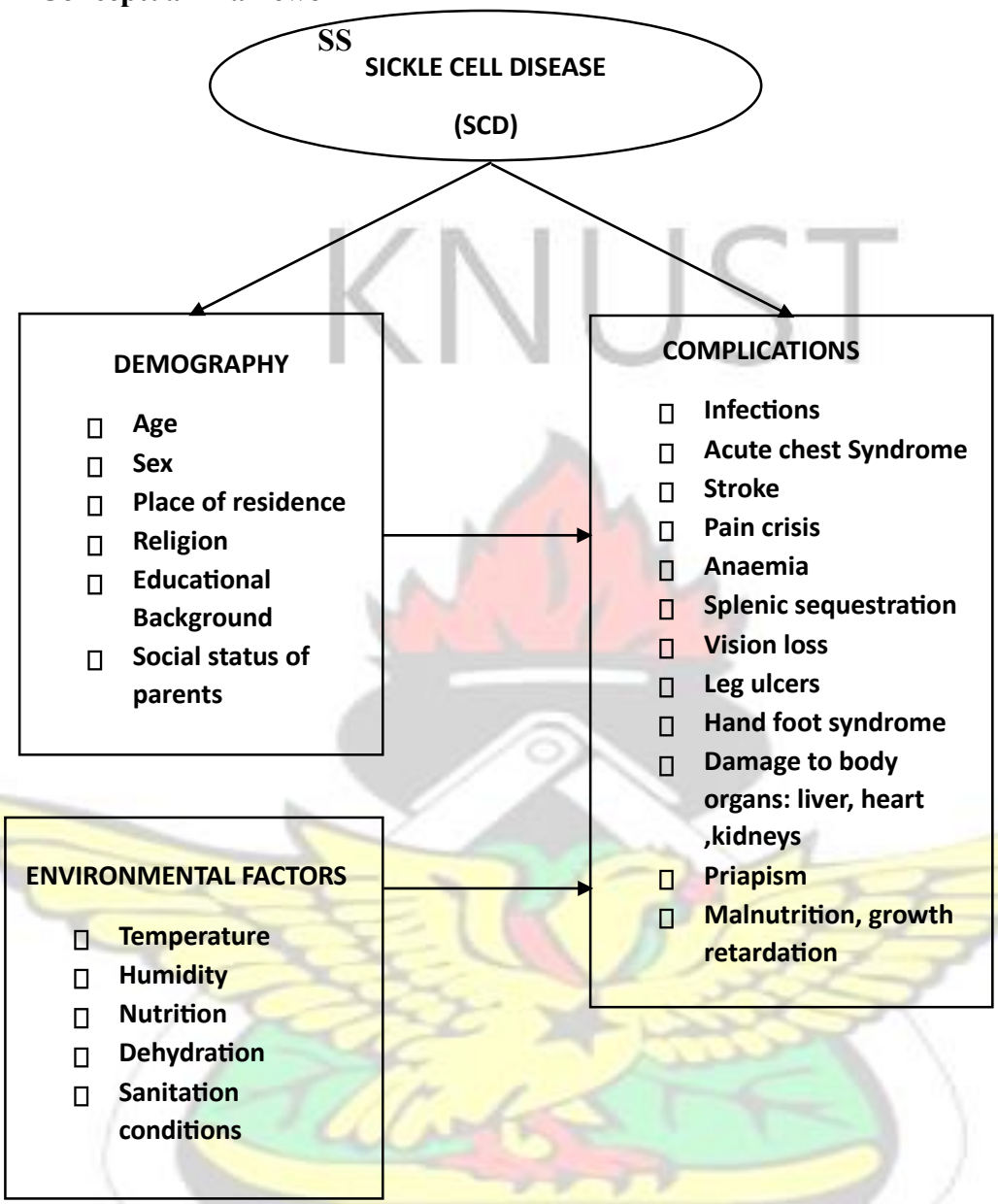
present study sought to throw light on the patterns of illness including comorbid conditions and how they predict outcomes of treatment.

1.3 Rationale for the study

Eck et al., (2004) as cited by Brown et al., (2013) made it clear that for effective planning by health officials for the care of children with sickle cell disease, it is important that knowledge of the patterns of the complications presented regularly to the hospital be known. Data on the most common conditions presented by sickle cell patients and the precipitating factors for these conditions will give more insight into the most severe of these conditions with the potential for mortality. Such data will assist in planning preventive strategies and resource allocation to reduce the effects on individuals and their families. Most of the studies done on patterns of illness of children with sickle cell disease have focussed on single complications such as patterns of crises alone or patterns of infections alone (Juwah et al., 2004), only a few studies have concentrated on the overall pattern of illness in children with sickle cell disease seeking care in a particular hospital (Ikefuna and Imodi, 2007).

In Ghana not many studies have been carried out in trying to determine the morbidity pattern of children with sickle cell disease. By carrying out this study at the KATH sickle cell clinic data is going to be made available on the common morbid conditions presented as well as triggering factors for these conditions at the sickle cell clinic which will help in planning preventive measures and identifying which areas to commit more resources and the kind of educational programmes to design for families and caregivers of these children. The outcome of which will be a reduction in the number of hospital visits and eventually reducing the burden of cost involved in regular hospital visits.

1.4 Conceptual Framework



Sickle Cell Disease (SCD) is known to be the most common genetic blood disorder in SubSaharan Africa (Weatherall & Clegg, 2001). People with SCD suffer from several

complications, some severe others mild and can occur suddenly (Ebert et al, 2010). Some of the complications include infections, pain crisis, acute chest syndrome, stroke, anaemia etc. SCD affects persons of all ages and both sexes, persons of high or low educational status, socio-economic status and persons with different religious beliefs are all affected by the disease.

Complications suffered by persons with SCD are mostly influenced by their age. Older age was associated with frequent episodes of pain (Darbari et al., 2012), sex, educational level and socio- economic status of persons with SCD also influenced how frequent and the kind of complications they will encounter. Educated people with a high socio- economic status who can afford housing in a clean environment, nutritious diet and access health care on a regular basis are more likely to have fewer complications than persons living in slums where infections are very common and cannot afford nutritious food and access healthcare on a regular basis.

Environmental factors such as exposure to cold weather and rainy seasons are known to increase pain episodes (Tewari and Rees, 2013, Mohanty and Mukherjee, 2002). During these periods persons with SCD get exposed to a lot of infections resulting in them having complications. A clean environment, good nutrition and access to public health measures as well as a high socio-economic status which may ensure persons with SCD have access to health care is relevant in determining the frequency of complications (Serjeant, 2013).

1.5 Research Questions

I. What proportion of SCD children accessing care at KATH have stroke?

II. What are the demographic characteristics of children with Sickle Cell Disease accessing care at KATH?

III. What are the common co- morbid conditions children with SCD present with to the

sickle cell clinic?

IV What are the predisposing factors accounting for the frequent presentation of complications to the hospital by children with SCD?

1.6 Research Hypothesis

I. There is no association between demographic factors of children with SCD and the development of crises.

II. Children with concomitant infections are more likely to suffer SCD crises.

1.7 Research Objectives

1.7.1 General Objectives

To identify the Patterns of Illness among children ≤14 years with SCD seen at KATH.

1.7.2 Specific Objectives

I. To estimate the proportion of SCD children who have stroke.

II. To determine the demographic characteristics of SCD children accessing care at KATH

III. To identify the common co-morbid conditions presented at KATH by SCD children.

IV To determine the predisposing factors accounting for the frequent presentation of complications by children with sickle cell disease.

1.8 Scope of the study

The study reviewed secondary data of children with Sickle Cell Disease (SCD) enrolled and assessing care at the Komfo Anokye Teaching Hospital (KATH) sickle cell clinic within the period 2012 – 2014. Data on the demographic characteristics, types of complications, number of visits and predisposing factors to the development of these complications among children

with SCD were extracted and analysed to determine the common co-morbid conditions, associated infections and the predisposing factors that trigger these complications among these children. A data abstraction form which contained all the required variables was used to collect data from case notes of children with SCD.

1.9 Organisation of Report

This thesis is organized into six chapters. Chapter one is the Introductory chapter comprising of Introduction, Background information, Problem statement, Rationale of study, conceptual framework, Hypothesis, Research questions, General objective, Specific objectives, Scope of the study, organization of report and assumptions of the study. Chapter two reviews relevant literature on the topics of the study. Chapter three examines the methodology used for the study, chapter four deals with presentation of the study results. Chapter five discusses the finding and the last chapter contains conclusions and recommendations

1.10 Assumptions of the Study

Source of data storage and retrieval reflect actual situation over the period under study.

CHAPTER TWO

2.0 LITERATURE REVIEW

Introduction

Inherited haemoglobin disorders (sickle-cell disorders and thalassemias) were formerly conditions of the tropics and subtropics but are currently worldwide due to migration (Weatherall and Clegg, 2001). Since these conditions can be controlled effectively by programmes aimed at treatment with carrier detection and genetic counselling, WHO has recommended the development of these programmes to reduce the occurrence of these disorders (World Health Assembly, 2006). Although over 700 structural haemoglobin variants have been identified the common ones include Hb S, Hb C, Hb E, Hb D etc. β thalassemia, α^0 thalassaemia. α^+ thalassemia includes heterozygous and homozygous α^+ thalassaemia. Sickle cell disorders include SS, SC, and S/β thalassaemia. Whereas thalassemia's include homozygous β thalassaemia, haemoglobin E/β thalassaemia,
homozygous α^0 thalassaemia, $\alpha^0/\ \alpha^+$ thalassaemia (haemogloerbin H disease).

Although all these disorders can be identified, sickle cell disorders are the most common that affects millions of people worldwide. It refers to a group of conditions characterised by the presence of haemoglobin S (HbS) and another abnormal haemoglobin. It is caused by a genetic abnormality in the gene for haemoglobin which results in the production of sickle haemoglobin. This sickle shaped red blood cells break apart easily causing anaemia. The damaged sickle red blood cells also clump together and stick to the walls of blood vessels blocking blood flow leading to severe pain and permanent damage to the brain, heart, kidneys, liver, bone and the spleen and a host of other complications. According to World
Health Organisation (WHO), 5.2% of the world's population and over 7% of pregnant women carry an abnormal haemoglobin gene (Modell and Dalison, 2008). In Africa, it is estimated that a total of 200,000 children are born annually with the condition (Diallo and Tchernia, 2002).

The common SCD disorders in Sub-Saharan Africa include homozygous HbSS disease (HbSS) commonly known as sickle cell anaemia (SCA) and HbSC disease.

Clinical manifestations of SCD usually begins in childhood, these complications are mainly divided into four categories: haemolysis and haematological complications, vaso-occlusion, infections and organ dysfunction (Makani et al., 2013). These complications are most often triggered by factors such as unfavourable climatic conditions such as cold and rainy weather (Mohanty and Mukherjee, 2002).These periods are associated with the spread of several infections which is an underlying cause for acute illness in persons with SCD.

For effective management of SCD and other haemoglobinopathies emphasis should be placed on new-born screening, anti-microbial prophylaxis, vaccination against infections, and training of healthcare workers, patients and caregivers (Ansong et al., 2013)

2.1 Demographic characteristics of children with SCD and other Haemoglobinopathies.

This refers to the characteristics of a population, as classified by age, sex, income etc. for research, sociological analysis etc. (Webster's College New World Dictionary, 2010). For the purposes of this study, demographic characteristics will look at the age, sex, as well as the place of residence of children with sickle cell disease since these indicators play a major role in determining the morbid conditions children with SCD may present to the various hospitals as well as the precipitating factors to some of the complications they present.

2.1.1 Age of children with SCD and other Haemoglobinopathies

Age has been identified as an important demographic characteristic in the study of children with SCD. A study by Jain et al, (2013) found infants making up 18% of the total population whereas children younger than three years made up 56% of the study population. In a different study, children <16 years who met the criteria for anaemic crises were selected for the study.

In this same study, reference was made to the fact that 30 000 Igbo preschool children suffer from sickle cell anaemia (Juwah et al., 2004), giving an indication of how common the disease is, among children between two to three years.

Another study Brown et al., (2013) in Nigeria used children between the ages 0-≤15 with majority of them falling within the age bracket 5-9 years (44%) an indication of how common SCD is among children of all ages.

Again children < 18 years were used in the study with the objective of determining morbidity among children with SCD during and after travels to a tropical area. The study however concluded that children travelling to tropical areas are more associated with high morbidity (Sommet et al., 2013). In a different study to assess pain episodes in children and adolescents with SCD, parents of children between the ages 6- 17 years completed a daily dairy about their children's pain response for 14 days. Results indicate that children with SCD usually experienced low levels of pain that were easily managed at home sometime without any medication (Gill et al., 2000)

2.1.2 Sex of Children with SCD and other Haemoglobinopathies

Sex refers to the biological differentiation between male and female.

A study to look at the morbidity patterns in hospitalised under five children with SCD in India identified a total of 85 children with 71% being boys' whiles the rest were females (Jain et al, 2013). The higher number of boys in this study is similar to another study (Kamble and Chaturvedi, 2000) in central India giving an impression of high prevalence of SCD in male children in certain communities in India.

In a different study carried out in Nigeria where sex, age and social class were the sociodemographic features used for selection of children for the study. Majority of the children (55.9%) were males, given an impression of more male children having SCD compared to females (Brown et al, 2013).

In the Health- Related quality of life study (Wrotniak et al., 2014) carried out in the U.S.A using the Child Health Questionnaire. Most of the children used for this study were males (57%) an indication that SCD is not only common in male children in some parts of Africa but also in some parts of the U.S.A. However, in Fung and colleagues study (2006), which sought to look at morbidity and mortality in chronically transfused subjects with thalassemia and sickle cell disease, majority (52%) of the subjects were females. One of the few studies where females have been identified to be in the majority. Another study carried out in Barbados to identify clinical findings associated with SCD in the population, majority (67%) of the participants who took part in the study were females which gives an indication that there are communities in the Caribbean where more females have SCD as compared to males

(Quimby et al., 2014).

2.1.3 Place of residence of children with SCD and other Haemoglobinopathies.

Place of residence refers to a place where a person resides over a period of time. In the Jain and colleagues (2013) study, one third of the children were residents of the Mahar community followed by the Kumbi (20%) Gondh (20%) and Teli (13%) an indication of how common SCD is in these communities in India. Another study also found children in the Igbo community of Nigeria having as many as 3000 children with Sickle cell anaemia (Juwah et al, 2004). A different study conducted in India also found that the prevalence of SCD to be highest in the state of Chhattisgarh (23%). Although the prevalence is high for tribal and scheduled caste populations like Kumi (55%) and Teli (53%) all belonging to the backward castes. These findings give an indication of how common SCD is, in most communities in India. These findings also give an impression of how common it is, to find children with SCD in most communities the world over (Patra et al., 2010).

2.2 Common co-morbid conditions presented by children with SCD

The clinical features of SCD in children may be severe or chronic, they cannot be predicted most often and can be life threatening (Stuart and Nagel, 2004). Some of these conditions include pain crises, anaemia, acute chest syndrome, leg ulcers etc. These conditions bring about a lot of suffering to these children and their parents.

Some of the common conditions children with SCD in an Australian paediatric population presented for hospitalisation were as follows occlusive crisis (70.8%) followed by infections (12.1%) and anaemic episodes. Making vaso- occlusive crisis the most common condition among children with SCD in that population (Teoh, et al., 2012) this is in line with a study in Nigeria which also identified vaso-occlusive crisis (61.5%) as the most common reported crisis, however in the Nigerian study, the second most common morbid condition presented was hyper- haemolytic crisis (16.7%) and then acute splenic sequestrations follows with 6.9%, the least common condition presented was the aplastic type 0.6% (Brown et al, 2013). This gives an indication of how common vaso-occlusive crisis is in children with SCD irrespective of age, sex, race or ethnicity. Again in the same study by Brown and colleagues out of the number that had the above complications 74.6% had associated infections, the most common ones being septicaemia (32.2%), malaria (28.2%), acute osteomyelitis (13.8%) and pneumonia (13.2%). Whereas the less common associated infections were urinary tract infection (6.9%), septic arthritis (5.7%), pharyngitis (2.9%) and chronic osteomyelitis. This result provides evidence of vaso-occlusive crisis accounting for majority of hospital admissions of SCD in developed countries. Whereas infections account for most admissions in developing countries (Fosdal and Woiner-Alexandrov, 2007).

In a different study to identify the incidence of serious bacterial infections in febrile children with SCD where serious bacterial infection was defined as bacteraemia, meningitis, urinary tract infection, osteomyelitis and pneumonia. Overall results indicated a 16% incidence rate of

bacterial infection among children used for the study. Pneumonia had the highest incidence rate of 13.8% followed by bacteraemia and urinary tract infections with incidence rate of 1.1% respectively (Bansil et al, 2013).These findings allowed the researchers to conclude that pneumonia is the common serious bacterial infection among the study participants.

In the Brazzaville study, infections (36.6%), vaso-occlusive crises (26.7%) and anaemic crisis (20.3%) were the three most dominant complications among children with SCD. The objective of the study was to assess the frequency and the nature of complications and prognosis of the disease in children suffering from sickle cell disease (Mabiala-Babela et al., 2005) and the results were similar to the study by (Brown et al., 2013) and also supports the assertion that infections account for most admissions in developing countries.

Another study carried out in India by Jain et al., (2013) to identify morbidity patterns in hospitalised under five children with SCD revealed that acute febrile illness (31%) was the most common morbid condition followed by severe anaemia (30%) and acute painful events (20%). Hand – Foot syndrome (11%), splenic sequestration crisis (4%), acute chest syndrome (3.3%) and stroke (0.6%) accounted for the rest.

In a different study to determine the main causes of hospitalisation, results indicated that blood transfusion, sickle cell crises (SCC) and infections were the most common causes of hospitalisation. This led the authors to conclude that non- compliance with medication increased patients' susceptibility to SCC and infections (Patel and Athavale, 2004).

Whereas the study by Akar and Adekile, (2008) with the objective of identifying the common causes of hospitalisation in Kuwait identified vaso- occlusive crisis (63.2%), acute splenic sequestration (9.1%), haemolytic crisis (8.8%) and acute chest syndrome (6.6%) as the common morbid conditions that resulted in hospitalisation of children with SCD. Stroke was seen in only one percent of children whiles acute osteomyelitis was also seen in just a percent of all the admissions.

Results from a study carried out to determine the prevalence of Airway Hyperactivity (AHR) in children with SCD who do not have asthma or any recent history of acute chest syndrome (ACS) revealed that, 29 (72.5%) of 40 children had a positive on the methane challenge test (MCT). Nine (31%) also reported with asthma like symptoms on questionnaire. This led the authors to conclude that there is a high prevalence of AHR that is not associated with asthma like symptoms in children with SCD (Shilo et al., 2015)

A study by Komba et al., (2009), to determine malaria as a cause of Morbidity and Mortality among children with homozygous sickle cell disease on the coast of Kenya, came to the conclusion that there was no evidence to support the risk of malaria being high among children with SCD than in children without SCD.

2.3 Proportion of Sickle Cell Disease Children with Stroke

Stroke is one of the complications children with SCD suffer. It can happen if sickle cells get stuck in a blood vessel and clog blood flow to the brain. About 10% of children with SCD will have a symptomatic stroke (CDC, 2013). Stroke can cause learning problems and lifelong disabilities. Several studies have looked at the link between stroke and SCD in children.

In a study by Earley and colleagues, (1998), Sickle Cell Disease was identified to have played a major role in ischemic stroke among children. Children in the age bracket 1 to 14 discharged from 46 hospitals in Central Maryland were used for this study. Eighteen of these children were diagnosed of ischemic infarction and 17 with intracerebral haemorrhage. The most common cause of ischemic stroke was Sickle Cell Disease (39%) leading to the conclusion that SCD plays a disproportionately high role in childhood stroke in the population surveyed.

In a similar study to evaluate trends in stroke hospitalisation among children (0 -17 years) and adults >17 years with SCD in the United States, results showed that paediatric patients with SCD co-morbid conditions constituted 8.7% in 1997 versus 4.8% in 2006 giving an indication

of a reduction in the number of cases in 2006 largely due to an increase in prophylactic red blood transfusion as against an increase in the cases among adults from
0.3% in 1997 to 0.5% in 2006 (Ovbiagele and Adams, 2012).

In a different study to identify the rates and risk factors for cerebrovascular accidents in SCD, results showed a prevalence rate of (4.1%) and an incidence of (0.61 per 100 patient-years) in sickle cell anaemia (SS) patients, however cerebrovascular accidents were also found to have occurred in all the common haemoglobin genotypes. The study also found the occurrence of infarctive CVA to be low in SS patients 20-29 years of age but higher in children and older patients (Ohene-Frimpong, 1997). In a related study conducted earlier by Balkaran et al., (1992), to identify stroke in a cohort of patients with homozygous SCD, results showed that stroke was identified in 17 of the 310 patients with homozygous SCD representing those who were followed from birth which represents an incidence of 7.8% by age 14 years. The common cause of stroke was cerebral infarction found among 15 of the 17 cases. Supporting the assertion of infarction being a common cause of stroke in children with SCD.

A different study carried out by (Quinn et al., 2013) with the objective of testing the hypothesis that acute cerebral ischemic events are frequent and potentially transient among children with SCD, results indicated that acute silent cerebral ischemic events were detected
1.3% of MRI (10 of 771) in 652 children with an incidence of 47.3 per 100 patient years. This led the researchers to conclude that children with SCA experience ongoing cerebral ischemia, sometimes reversible far more frequently than previously recognised. Other studies have also identified a very low percentage of stroke among children with SCD. In Jain and colleagues study, (2013) stroke was identified in only 0.6% of the children less than 5 years with SCD and 1% identified in the study by (Akar and Adekile, 2008), giving an indication of an overall low percentage of stroke in SCD children.

A study conducted in France on SCD documented a 6.7% prevalence of clinical overt stroke and a 15% prevalence of silence infarcts in sickle cell anaemia (SCA) patients with African descent. This finding goes to support the findings of a number of studies on stroke where the prevalence was less than 10% among children with SCD (Bernaudin et al., 2000)

2.4 Predisposing factors to morbid conditions in Children with SCD

A lot of factors account for how severe and frequent children with SCD may suffer from complications. These factors are largely genetic and environmental. Genetic factors such as high levels of Fetal Haemoglobin at birth is said to protect the child against vaso-occlusive and haemolysis in the first few months of the child's life. The type of Sickle Cell trait a child has also has a great influence on how regular he may present with complications. Environmental factors such as climatic conditions, exposure to Streptococcus pneumonia or parvovirus B19 and other infections like salmonella can cause severe complications in the child. Malaria continues to be a major determinant of morbidity and mortality among children with SCD. Other important factors are nutrition, access to public health measures such as immunization, and socioeconomic status, which may determine the families' access to communication, transport, and medical care are very important in determining the severity and how regular children with SCD will visit the hospital (Serjeant, 2013).

A study in Egypt was conducted to identify the predictive factors for acute painful crises in children with SCD to determine preventive strategies. Results indicated that pain rate was very high in children with Haemoglobin SS especially those with an early onset of dactylitis. However the top 3 predictors of SCD severity were identified in descending order as genotype, basal haemoglobin level and dactylitis (Al-Haggar et al, 2006).

In a different study by Silva et al., (2015), leucocytosis and dactylitis in the first year of life was also identified as a predictor of severity of SCD in children, while the presence of

αthalassemia is known to be a protective factor. They identified a statistically significant association between leukocytes >15 000/μL and a higher number of hospitalizations (P < 0.001) and chronic complications of the disease (P = 0.035). The occurrence of dactylitis in first year of life was also significantly associated with a higher number of hospitalizations (P = 0.004) and chronic complications (P = 0.018). The presence of α-thalassemia was associated with a lower number of chronic complications (P = 0.036).

Another study carried out to identify risk factors for headaches and migraines among children with SCD came to the conclusion that low haemoglobin levels were associated with recurrent headaches and migraines (Dowling et al., 2014).

Darbari et al., (2012),conducted a study to determine the risk factors associated with severe frequent vaso- occlusive crises in a contemporary paediatric cohort of patients with Sickle Cell Anaemia, results showed that frequent pain episodes were associated with older age, αthalassemia trait, iron overload and higher levels of haemoglobin giving an indication of the varied nature of factors that can trigger complications.

In a separate study conducted to identify and compare factors that affect the clinical outcomes of Sickle Cell Anaemia in patients 11 -30 years in Nigeria and the U.S.A, findings revealed that higher values of Body Mass Index (BMI) and Blood Pressure (BP) in Chicago Sickle Cell Anaemia patients may contribute to an increased risk of stroke, whereas lower pneumococcal vaccination and Hydroxyuria therapy rates in Ibadan patients highlights the need for more improved vaccination coverage (Akingbola et al., 2014) to help reduce the frequency of visits to the hospital. These findings suggest that nutrition and access to public health interventions such as immunisations are important in reducing morbid conditions in children with SCD.

The relevance of climatic factors as predictors of acute pain has been known over 30 years and several studies have shown an increase in pain episodes in cold and rainy seasons although patterns vary (Tewari and Rees, 2013). In India it is known that the highest incidence of vaso-

occlusive crisis occurs during the rainy season followed by winter (Mohanty and Mukherjee, 2002). During the rainy seasons children with SCD are exposed to a lot of infections and infections have been identified as a common underlying cause for acute illness in SCD in India (Jain et al., 2013). In a different study admissions have been associated with increased wind speed and low humidity but there was no association with rainfall or barometric pressure and temperature (Jones et al, 2005).

An analysis of environmental factors on a large population of people with SCD in South London revealed that windy weather and low humidity was associated with increased hospital admissions. Again air quality was also identified to be associated with acute complications of SCD with high levels of ozone (O3) responsible for more episodes of acute pain. On the contrary high levels of Carbon Monoxide (CO) and Nitrogen Oxide (NO) were linked with fewer hospital visits (Mittal et al, 2009).

In a different study to determine whether surgery was a risk factor in the development of ACS among children with SCD, results revealed that abdominal surgery was associated with a higher risk (20%) of developing ACS. Laparoscopy was identified not to decrease the occurrence of ACS as compared with open approach. This gives an impression of the varied nature of triggering factors for the development of crises in children with SCD.

Another study which sought to determine the association between the type of opioids selected for the treatment of pain crisis and the development of ACS, results showed that of the total of 37 (21%) episodes of ACS, 26 (29%) were in the morphin group whereas 11(12%) were in the Nubian group ($P<0.01$). This led the researchers to conclude that developing ACS during pain episodes is as a result of varied factors however the type of opioid selected may increase the rate (Buchanan et al, 2005).

Another study which sought to look at the complications associated with human parvovirus B19 infection in children with SCD identified the following clinical events: fever (89.7%), pain

(61.8%), acute splenic sequestration (19.1%) and acute chest syndrome (11.8%). Pain, fever and acute splenic sequestration were more commonly associated with acute HPV B19 infection in children with SCD than those without the disease (Smith- Whitley et al., 2004).

Results from a study by Serjeant and colleagues (1994), to identify the clinical features as well as perceived predisposing factors for painful crises of homozygous SCD revealed that of the perceived predisposing factors, skin cooling occurred in 34%, emotional stress in 10%, physical exertion in 7% and pregnancy in 5% of women of child-bearing age. This makes it obvious that a lot of factors are perceived to trigger pain episodes in people with SCD.

2.5 Patterns of Illness among children with SCD

Patterns of illness refer to the different kinds of conditions that are presented to various health centres by children with SCD. Several studies have been done by various authors to identify different types of complications presented to hospitals by children with SCD.

A study was carried out by (Tarer et al., 2006), on the patterns and prevalence of acute clinical events on children with sickle cell anaemia in Guadeloupean. Results indicated that painful crisis and acute chest syndrome were the common complications affecting 65.4% and 58.8% of the patients, respectively. The frequency of acute anaemia was 49.7% (acute splenic sequestration 24.8%; acute aplastic anaemia 15.0%). Prevalence of septicaemia-meningitis and osteomyelitis were 15.7% and 16.3%, respectively. A higher incidence of infections, painful crises and acute anaemia was detected in patients who developed ACS.

In a similar study carried out in Brazil to look at morbidity and mortality among people with SCD in two different hospitals from 1998 to 2007. Findings showed that the most frequent reasons for visits to these hospitals were afebrile pain episodes (69.1% and 25.3% respectively). The epidemiological profile of the study showed a predominance of children, and young adults, women and the genotype SS (Paulo Juliano Roberto et al., 2010).

Another study which looked at the clinical events in the first decade in a cohort of infant with

SCD enrolled 694 infants at less than 6 months of age and information about nature and frequency of complications presented was collected over a ten year period. Painful crises and acute chest syndrome were the most common sickle cell related events in homozygous sickle cell anaemia (SS), haemoglobin SC disease (SC) and Sβ thalassemia patients with overall incidence in SS patients 32.4 and 24.5 cases per 100 person-years, respectively. Bacteraemia was also identified to have occurred in most SS children under 4 years of age and in SC children less than 2 years of age (Blood, 1995).

In the study by Brozovic and colleagues (1987), all acute admissions of patients who lived in the London Borough of Brent and attending a particular hospital were analysed for a period of one year. Sixty three of the 211 who were followed up required 161 admissions during the year. Most admissions (126) were for the 42 patients with homozygous sickle cell disease; 147 (91%) were for vaso-occlusive episodes, 142 of which were for painful crises, three for cerebrovascular accidents, and two for renal papillary necrosis. This findings and that of the other studies above gives a clear picture of vaso- occlusive crises being the most common among SCD patients.

Results from a different study by Jain et al., (2013) which looked at morbidity patterns among under 5 children with SCD identified acute febrile illness (31%) as the most common morbid condition followed by severe anaemia (30%) and acute painful events (20%).One of the few studies contradicting the findings of other studies where painful episodes are the common crises presented.

In the Quimby and colleagues study (2014) to assess the clinical findings associated with homozygous SCD in the Barbadian population, results indicated that 94% had ever had painful crisis. In the previous year, 44% of the participants had at least one crisis. Chronic leg ulcer was prevalent 27%, whereas forty two persons had urinalysis and out of that number 44% were

diagnosed with albuminuria. This results makes it clear of the varied nature of clinical complications persons with SCD present with.

CHAPTER THREE

3.0 METHODOLOGY

3.1 Study Design

This was cross-sectional that involved use of secondary data from the sickle cell clinic at KATH for a period of three years (2012-2014).

3.2 Data Collection Techniques and Tools

Data were extracted for a three year period. Data extracted included demographic details, haemoglobin genotype, types of crises, associated infections, diagnosis and number of visits within the period. Demographic details collected were age, sex and place of residence. Data were extracted onto a well-designed abstraction form with the required indicators needed to help answer our research questions.

3.3 Profile of study Area

The study was conducted at the Sickle Cell Clinic of the Komfo Anokye Teaching Hospital (KATH), Kumasi, Ghana. Komfo Anokye Teaching Hospital is located in the vibrant and culturally rich city of Kumasi, (Bantama sub-metro) the regional capital of Ashanti, with a population of about 4.7 million (2010 population census).

The Ashanti Region occupies a total land area of 24, 389 square kilometres representing 10.2% of the total area of Ghana making it the third largest region after Northern and Brong Ahafo Regions. It has a population density of 148.1 persons per square kilometer which also makes it the third after Greater Accra and Central Regions. It constitutes 14.8% of all the total Akan population in the country. There are 27 administrative districts in the Region, including the Bantama Sub-Metropolis which forms part of the Kumasi Metropolitan Assembly.

The geographical location of KATH, the nature of road network in the country, as well as the commercial nature of the city makes the 1,200-bed capacity teaching hospital accessible to all

the areas that share boundaries with Ashanti Region even others further away. The hospital therefore receives referrals from almost seven other regions including the three Northern regions.

Historically, the origin of the hospital dates back to 1940, where it was located on the hill overlooking Bantama Township. It was then called, African and European Hospital and as the name implies, the African section took care of Africans whilst the European side provided treatment for the Europeans. However, on some occasions, high-ranking African Government Officials were given treatment at the Europeans section.

As the population in Kumasi increased, the need to construct a new hospital arose in 1952. The Europeans section was transferred to Kwadaso Military Quarters to make way for the project to begin. Kumasi Central Hospital was the name of the new hospital complex which was completed around 1954/55. The name was later changed to Komfo Anokye Teaching Hospital in honour and memory of the legendary fetish priest, Komfo Anokye.

It became a teaching hospital for the training of medical students from the Kwame Nkrumah University of Science and Technology (KNUST) in 1975. It is the second largest Teaching Hospital in the country, and the only tertiary institution in the Ashanti Region. It serves as a major referral centre in the northern sector of Ghana. Presently it is a training centre for Ghana Post Graduate College of Physicians and Surgeons. The hospital also provides training for nurses and midwives from Kumasi Nurses and Midwifery Training College and nurses from other nurses training colleges in the metropolis as well as Pharmacy and Medical laboratory scientist students from the KNUST.

The hospital has (12) clinical directorates, and (2) non- clinical directorates namely surgery, obstetrics and gynaecology, child health, medicine, polyclinic, diagnostics, emergency medicine, traumatology and Orthopaedics, oncology, anaesthesia and intensive care, EENT, oral health, domestics and Technical Services. The hospital has a staff population of 3,909 who

fall under these categories, Doctors (9.4%), Top Management (0.2%), nurses and midwives (42.2%), Physician Assistants (1.3%), pharmacist and pharmacy technicians (3.8%), Administration and Finance (6.6%), clinical support (10.9%) and Allied Health (5.6%) (KATH Annual Report, 2013).

KATH Sickle Cell Clinic falls under the directorate of child health and it is one of the subspecialist clinics of the directorate with the others being the Paediatric HIV Clinic, Asthma clinic, Cardiology, renal clinic etc. (KATH Annual Report, 2013). The clinic started operating in 1992 and sees an average of 5000 children from age 0 -14 years per annum. Clinic days are Mondays where an average of 170 children are seen. These include both new and old patients. The clinic's staff strength are as follows, 7 resident Doctors, and about 10 house officers who come there on clinic days, 4 nurses, 3 records officers and 2 data entry clerks.

3.4 Study Population

The study population were children ≤ 14 years with Sickle Cell Disease who were enrolled and who accessed care at KATH from 2012 to 2014.

3.5 Study Variables

Dependent variable: Vaso-occlusive crisis, Haemolytic crisis, Acute chest Syndrome, Stroke, Splenic sequestration, Vision loss, Leg ulcers, Infections, Hand- foot- syndrome, Damage to body organs: (liver, heart, kidneys,) Priapism, Malnutrition, growth retardation etc.

Independent variables: Age, sex, place of residence and haemoglobin genotype, educational background and religion.

Table 1: Definition of variables for the study

Variables	Definition	Indicator	Measurement	Objective Addressed

Prevalence of SCD children with Stroke	Proportion of children who had stroke in the period 20122014	Number of children	Discrete	One
Demographic Characteristics of Children with SCD.				Two
Age	Age in months as at last birth date	Categories of age 0-4 5- 9 10 – 14	Ordinal	
Place of Residence	Place one stayed over the period of time	Urban Rural	Binary	
Sex	Biological differentiation of male and female	Male Female	Binary	
Haemoglobin Genotype	Types of haemoglobin	Hb SS, Hb SC, Hb S/β°- Thalassemia	Nominal	
Co-morbid Conditions	Any disease sign/symptom associated with children with SCD			Three
Vaso- occlusive crisis	Mild to severe pain which starts suddenly and last any length of time	Yes No	Binary	
Haemolytic crises	Rapid destruction of large numbers of red blood cells	Yes No	Binary	

Hand-footsyndrome	Swelling of hands and feet in infants and young children.	Yes No	Binary	
Acute Chest Syndrome	Chest pains, coughing, difficulty in breathing and fever.	Yes No	Binary	
Splenic Sequestration	Spleen enlargement due to trapping of sickle cells in it.	Yes No	Binary	
Stroke	Sickle cells getting stuck in blood vessels and blocking blood flow.	Yes No	Binary	
Malaria	Parasite causing blood disease transmitted through the bite of the anopheles mosquito	Yes No	Binary	
Septicaemia	Infection in which large amounts of bacteria are present in the blood	Yes No	Binary	
Pneumonia	Lung inflammation caused by bacteria or viral infection	Yes No	Binary	

Predisposing Factors for development of SCD complications				Four
Age	Age in months as at last birth date.	0-4 5- 9 10 – 14	Ordinal	
Place of Residence	Place one stayed over the period of time	Urban Rural	Binary	
Sex	Biological differentiation of male and female	Male Female	Binary	
Haemoglobin Genotype	Types of haemoglobin	Hb SS, Hb SC, Hb S/β°- Thalassemia	Nominal	
Malaria	Parasite causing blood disease transmitted through the bite of the anopheles mosquito	Yes No	Binary	
Septicaemia	Infection in which large amounts of bacteria are present in the blood	Yes No	Binary	
Pneumonia	Lung inflammation caused by bacteria or viral infection	Yes No	Binary	

3.6 Sample Size

Assuming a prevalence of 50% of the most common SCD crises (vaso- occlusive) to occur within the population, we would require 234 patient folders over the period of three years. Assuming a margin of error of 5% and 95% confidence interval and assumed non-completion rate of 10% using Epi Info statistics calculator.

On a probability proportion to size based on previous reports from patient folders of the three years under study we weighted the samples to reflect anticipated yield per year giving us 70, 80 and 84 folders for 2012, 2013 and 2014 respectively.

3.7 Sampling technique

Folders eligible for extraction were those of children ≤ 14 years with SCD enrolled into care in the period under study. They were 601 for the three year period. These were spread per year as follows: 2012, (181 patients), 2013, (208 patients) and 2014 (212 patients) respectively. All folders of children enrolled in each year were numbered from one to the last enrolment number. On the basis of probability proportional (PPS) to size method I estimated the proportion each year contributed to the total sampling frame by dividing the total enrolment in each year by the total population for the three years. On this basis the first year which is 2012 contributed 30% to the total population, 2013 contributed 34% and 2014 contributed 36% to the total population. This translated into a sample size of 70 folders for 2012, 80 folders for 2013 and 84 folders for 2014 which added up to the required sample size of 234.

Using the systematic sampling method, the sampling interval for the first year which is 2012 was calculated by dividing the total enrolled for that year which is 181 by the sample size which is 70 to arrive at a sampling interval of 2.58, which was rounded up to 3. The first folder was randomly picked using the lottery method. Folder number 2 was picked, which became the starting point, subsequently picking folders 5, 8, 11, 14…… until the 70th folder

eligible for extraction was picked for 2012. The same method was used to select folders for 2013 and 2014.

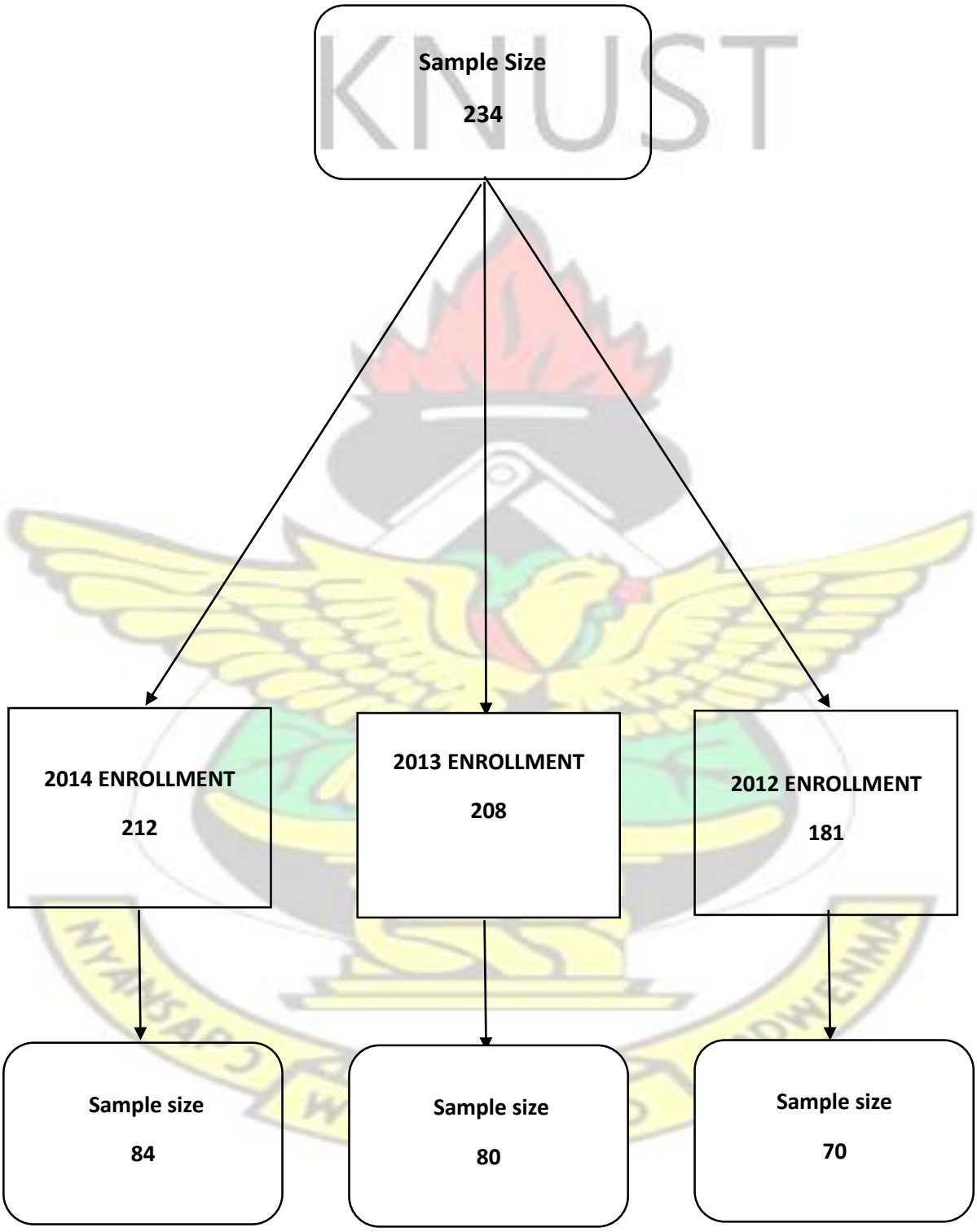


Figure 1: Sampling Technique Flow Chart

The following were considered in selecting eligible folders for extraction:

- Folders of children ≤14 years of age enrolled into care at the Sickle Cell Clinic within the period.
- Folders of children with at least one follow-up visit were considered for extraction.
- Folders of children more than >14 years of age were considered ineligible
- Folders of children with no follow-up visits were also considered ineligible.

Folders of children ≤14 years of age which were selected and did not meet the eligibility criteria of at least one follow-up visit were filed and the next folder in the predetermined pattern picked until the sample size was arrived at.

3.7 Pre- testing

Pre-testing of data collection tool was done at the Sickle Cell Clinic of the Maternal and Child Health Hospital (MCHH), Pampaso with 10 folders on one clinic day to test for the validity and reliability of the data collection tools. The pretesting ensured that a field called "others specify" was added to the abstraction form to make it possible to capture certain conditions which were not included in the tick option field. This helped to capture all the conditions presented by children with SCD and not only the major crises.

3.8 Data Management and Analysis

Five research assistants and one data entry clerk were identified and trained to help extract data onto the abstraction form. Data were then entered into the Epi Info Software version 7.1.6 for analysis. Data verification was carried out by the principal investigator by randomly selecting a folder which had already been extracted and comparing the information in it with the one on the abstraction form before it was entered into the database.

Data extracted from patients' folders were anonymised to protect their identity.
Hard copies of data were kept in a safe under lock and key and soft copies were password protected on the laptop of the principal investigator and also backed up on an external drive

which was kept in the same locker to ensure that only the researcher and his assistants had access to them when necessary.

Epi Info software version 7.6.1 was then used to compute for means and standard deviations for continuous variables and also generate frequency tables for categorical variables while stata software version 11 was used to test for hypothesis. This was done by first conducting a univariate analysis for predisposing factors such as age, sex, haemoglobin genotype, place of residence and infections such as malaria, septicaemia and pneumonia against the major crises to determine whether there was a relation. Those that were significant were then considered for further analysis. A multivariate analysis using logistic regression was then carried out to determine their statistical significance as well as the computation of odds ratios. Demographic variables such as age, sex, place of residence and haemoglobin genotype and infections such as malaria, septicaemia and pneumonia were all adjusted for. Categorical variables were presented in frequency tables and pie charts whereas outputs from logistic regression analyses were presented in tables.

3.9 Ethical Considerations

Ethical approval to carry out this study was giving by the Kwame Nkrumah University of Science and Technology Ethical Board. Approval to go on with the study at the Komfo Anokye Teaching Hospital (KATH) was granted by the Research and Development Unit of the hospital. Support and cooperation came from the Doctor In-charge and staff of the sickle cell clinic.

CHAPTER FOUR

4.0 RESULTS

Proportion of children with stroke

Three children, (one male and two females) (1.3%) were diagnosed with stroke. Their ages ranged from 4 – 8.5 years with a mean age of 6.8 years. Two of the children were of the haemoglobin genotype SS and that for the last child was not written in the patient's folder. All the cases of stroke were of the ischemic type, cerebral infarction was identified as the cause of stroke in one of the children and one was also identified to have been caused by spinal artery thrombosis. Two of the children had right hemiparesis as a result of the stroke. There were also co-morbid conditions of haemolytic crisis and pain episodes in two of the children. There were no associations found between the development of stroke and the predisposing factors analysed using logistic regression. However, children in the age group 5-9 were at a 1.3 times higher risk (AOR= 1.3, 95% CI. 0.3 – 43.1: p-value=0.852) of developing stroke, although not statistically significant.

4.1 Demographic Factors

Table 2: Demographic Characteristics of children with SCD

Variables	Frequency (n)	Percentages (%)
Age (years)		
0 – 4	108	46.2
5 – 9	81	34.6
10 – 14	45	19.2
Total	**234**	**100.0**
Sex		
Male	129	55.1
Female	105	44.9
Total	**234**	**100.0**
Place of Residence		
Urban	146	62.4
Rural	88	37.6
Total	**234**	**100.0**
Educational Background		
Nil	49	20.9
Crèche	25	10.7
Nursery/Kindergarten	61	26.1
Primary School	82	35.1
Junior High School	8	3.4
Not stated	9	3.8
Total	**234**	**100.0**
Religion		

Christian	191	81.6
Muslim	22	9.4
Not Stated	21	9.0
Total	**234**	**100.0**
Haemoglobin Genotype		
Hb SS	166	70.9
Hb SC	52	22.2
Hb S/β°- Thalassemia	1	0.4
Not Stated	15	6.5
Total	**234**	**100.0**

The ages of the children ranged between 4 months to 14 years with a mean age of 5.7 years and a standard deviation of 3.9 years. Majority of the children, 108 (46.2%) were within the age group 0 – 4 years. The proportion of male population was (129) 55.1%. Most of the children, 163 (62.4%) resided in urban areas. About 35% making up the majority were in primary school while the dominant religion among them was the Christian, practiced by 81.6% of children and their families. The common haemoglobin genotype was SS, 166 (70.9%) with S/β°- Thalassemia being the least common.

4.2 Crises presented by Children with SCD

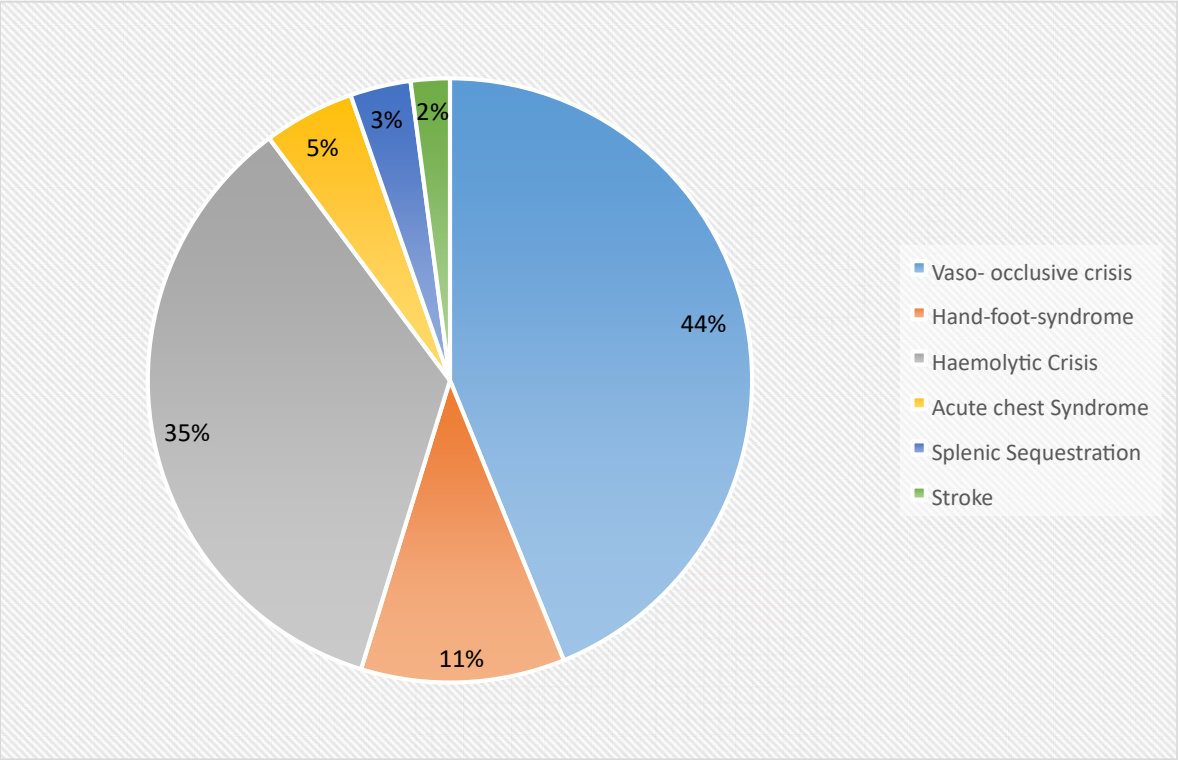


Figure 2: Proportion of common crises presented by children with SCD

A total of 1091 visits were made by 234 children to the sickle cell clinic during the period under study. Out of that 471 (43.2%) were routine visits with patients presenting with no complaints. Six percent of children made 11 visits to the clinic, which is the highest number of visits to the clinic by a patient within the study period. Several forms of crises were presented by 199 (85%) children during the 620 visits to the clinic with the most presented condition being vaso-occlusive crisis, (44%) followed by haemolytic crises (35%) while the least was stroke (2%)

4.3 Infections and other complications presented by children with SCD

Table 3: Description of Infections and Complications

Infections and other complications	**Number of events (n) %**	**Number of children (n)**
Septic Arthritis	8 (1.9)	6
Malaria	84 (20.8)	65
Meningitis	1 (0.2)	1
Pneumonia/Bronchopneumonia	14 (3.4)	14
Respiratory Tract Infection	47 (11.6)	38
Urinary Tract Infection	13 (3.2)	9
Fever/ Febrile illness	81 (20.1)	63
Parotitis	2 (0.5)	2
Avascular necrosis of femoral head	9 (2.2)	9
Chronic Osteomyelitis	20 (4.9)	18
Jaundice	54 (13.4)	49
Sepsis/Septicaemia	19 (4.8)	16
Liver disease	5 (1.3)	5
Priapism	3 (0.7)	3
Others	43 (10.7)	34
Total	**403 (100.0)**	**332**

*Multiple responses

Of the 199 children who presented with different kinds of crises, 165 (83%) had associated infections with the most common being malaria 84 (20.8%) and the least was meningitis 1 (0.3%).Other non-infectious complications presented included fever/febrile illness 81(20.1%), avascular necrosis of femoral head 9 (2.2%) and liver disease 5(1.3%).

4.4 Children presenting with co-morbid crises

Table 4: Frequency and age distribution of children with Co-morbid Conditions

Type of Crises	(N) %	Age Range (years)	Mean Age (SD)
Haemolytic crisis + Splenic Sequestration	3 (7.5)	2.5 – 8	5.5 (2.8)
Haemolytic crisis + Vaso-occlusive crises	12 (30)	2 – 12	7.4 (3.1)
Haemolytic crisis + Hand-foot-syndrome	6 (15)	1 – 7	2.7 (2.4)
Vaso –occlusive crisis + Hand-footsyndrome	10 (25)	0.8 – 4	2.1 (1.2)
Vaso –occlusive crisis + Acute chest syndrome	3 (7.5)	2– 14	8.3 (6.0)
Vaso – occlusive crisis + Splenic Sequestration	1 (2.5)	4	4
Haemolytic crisis + stroke	2 (5.0)	8	8
Vaso –occlusive crisis + stroke	1 (2.5)	4	4
Hand-foot-syndrome +Haemolytic crisis + Vaso-occlusive crisis	1 (2.5)	2	2
Hand-foot-syndrome + Vaso-occlusive crisis +Acute chest syndrome	1 (2.5)	6	6
Total	40 (100.0)		

SD =Standard Deviation

Forty children presented with Co-morbid crises with the most common being Haemolytic and Vaso-occlusive crises 12 (30%).The ages of the children who presented with these crises ranged

from 0.8 – 14 years. Males were dominant, 25 (63%). Haemoglobin genotype of the children were 33 (83%) for SS and 5 (13%) for SC while 2 (4%) were unknown

4.5 Association between predisposing factors and development of crises

Table 5: (a) Predisposing factors for the development of Vaso-occlusive Crises

Predisposing factors	Adjusted Odds Ratio	95% CI	P- value
Age			
- 0-4 years	1.00		
- 5-9 years	1.82	0.91- 3.65	0.091
- 10 – 14 years	4.87	2.16 – 10.99	0.001*
Sex			
- Female	1.00		
- Male	0.65	0.34 – 1.19	0.165
Haemoglobin Genotype			
- SC	1.00		
- SS	1.40	0.69 – 2.86	0.347
Place of Residence			
- Rural	1.00		
- Urban	0.58	0.31 -1.08	0.090
Infections			
- Malaria	4.71	1.60 – 13.86	0.005*
- Septicaemia	3.79	0.49 – 29.06	0.199
- Pneumonia	0.83	0.07 – 9.28	0.878

(*) = value is less than 0.05

A multivariate analysis conducted on the predisposing factors for the development of pain episodes found a strong statistical association between the age group 10-14 compared to the age group 0- 4 years. Being in this age group increased a child's chances of developing pain episodes by 4.8 times (AOR= 4.87, 95% CI 2.16- 10.99: p-value < 0.001). Sex had no statistical

association with pain episodes but the odds of having pain episodes was lower in males Having haemoglobin genotype SS increased the odds of experiencing pain episodes by 1.40 times (AOR= 1.40, 95% CI. 0.69 – 2.86:p-value=0.347) but not statistically significant. There was no significant relationship between place of residence and developing pain episodes. Having the malaria parasite also had a strong statistical association with the development of pain episodes and having malaria parasites increased a child's chances of developing pain episodes by 4.71 times (AOR= 4.71, 95% CI. 1.60- 13.86: p-value= 0.005) as compared to not having the malaria parasite. Though septicaemia had no statistically significant association with developing pain episodes, having septicaemia increased the chances of developing pain episodes 3.79 fold (AOR=3.79, 95% CI. (0.49 – 29.06: pvalue=0.199).

Table 6: (b) Predisposing factors for the development of haemolytic crises.

Predisposing factors	Adjusted Odds Ratio	95% CI	P- value
Age			
- 0-4 years	1.00		
- 5-9 years	2.23	1.04 – 4.75	0.037*
- 10 – 14 years	0.59	0.19 – 1.81	0.363
Sex			
- Female	1.00		
- Male	2.32	1.09 – 4.91	0.027*
Haemoglobin Genotype			
- SC	1.00		
- SS	1.35	0.57 – 3.17	0.484
Place of Residence			
- Rural	1.00		
- Urban	0.53	0.26 – 1.08	0.084
Infections			
- Malaria	1.67	0.51 – 5.43	0.389
- Septicaemia	2.99	0.38 – 23.00	0.292

(*) = value is less than 0.05

A multivariate analysis conducted on the predisposing factors for the development of haemolytic crises found a statistically significant association between the age group 5- 9 years (AOR=2.23, 95% CI, 1.04 – 4.75:p-value=0.037) and haemolytic crisis development. However if a child is within the age group 10-14 years his/her chances of developing haemolytic crises is lowered by 0.59 times (AOR=0.59, 95% CI, 0.19 – 1.81: p-value=0.363) comparing it to the age group 0-4. Sex had a statistically significant association with haemolytic crises and being male increased the chances of developing haemolytic crises 2.32 times (AOR= 2.32, 95% CI, 1.09- 4.91: p-value = 0.027). Haemoglobin genotype had no significant association with developing haemolytic crisis, however having haemoglobin type SS increased the chances of developing haemolytic crises 1.35 times (AOR= 1.35, 95% CI, 0.57 – 3.17: p-value=0.484). Malaria and septicaemia had no statistically significant association with the development of haemolytic crisis, however having the malaria parasite increases a child's risk by 1.67 times (AOR= 1.67, 95% CI, 0.51 – 5.43: p-value=0.389) of developing haemolytic crisis. Whereas having septicaemia also increases a child's risk of developing haemolytic crisis by 2.99 times (AOR= 2.99, 95% CI, 0.38 – 23.00: pvalue=0.292).

Table 7: (c) Predisposing factors for the development of hand-foot- syndrome.

Predisposing factors	Adjusted Odds Ratio	95% CI	P- value

Age			
- 0-4 years	0.09	0.02 – 0.41	0.002*
- 5-9 years	1.00		
Sex			
- Female	1.00		
- Male	0.75	0.29 – 1.94	0.555
Haemoglobin Genotype			
- SC	1.00		
- SS	3.08	0.64 – 14.76	0.158
Place of Residence			
- Rural	1.00		
- Urban	0.46	0.17 – 1.21	0.118
Infections			
- Malaria	1.37	0.25 – 7.25	0.709
- Septicaemia	1.35	0.10 – 16.85	0.814

(*) = value is less than 0.05

A multivariate analysis conducted on the predisposing factors for the development of handfoot-syndrome found a statistically significant association between the age group 0-4 years and the development of hand-foot-syndrome. (AOR= 0.09, 95% CI, 0.02 – 0.41: p-value= 0.002). Having haemoglobin genotype SS increased the chances of developing hand-footsyndrome by 3.08 times (AOR= 3.08, 95% CI, 0.64 – 14.76: p-value=0.158) as compared to having haemoglobin genotype SC. Though there were no association between infections such as malaria and septicaemia, and developing hand-foot-syndrome, having septicaemia increased a child's chances of developing hand-foot-syndrome by 1.35 times (AOR= 1.35,

95% CI, 0.10 – 16.85: p-value=0.814).Whereas having the malaria parasite also increases a child's odds of developing hand-foot-syndrome by 1.37 times (AOR=1.37, 95% CI, 0.25-

7.25:P-value=0.709).

Table 8: (d) Predisposing factors for the development of splenic sequestration.

Predisposing factors	Adjusted Odds Ratio	95% CI	P- value
Age			
- 5-9 years	1.06	0.09 – 12.11	0.962
- 10 – 14 years	1.00		
Sex			
- Female	1.00		
- Male	0.50	0.04 – 5.81	0.586
Place of Residence			
- Rural	1.00		
- Urban	1.41	0.12 – 16.27	0.779

(*) = value is less than 0.05

There was no statistically significant association between predisposing factors and developing splenic sequestration. However, if a child was within the age group 5-9 years the chances of him/ her developing splenic sequestration is increased by 1.06 times (AOR= 1.06, 95% CI, 0.09 – 12.11:p-value=0.962) as compared to the age group 10- 14 years. Residing in an urban area also increases the chances of developing splenic sequestration by 1.41 folds (AOR= 1.41, 95% CI, 0.12 – 16.27: p-value=0.779)

CHAPTER FIVE

5.0 DISCUSSION

5.1 Demographic Characteristics and Development of SCD

The study carried out revealed that SCD was responsible for the frequent presentation of complications by children to the Sickle cell clinic of KATH. Males were identified to be dominant in this current study, constituting 55.1%. This gives an indication of more male children affected by SCD than their female counterparts similar to the study by (Kamble and Chaturvedi, 2000, Brown et al., 2013) but a few studies have found females to be dominant (Fung et al., 2006, Quimby et al., 2014). However in most studies males are in the majority. The reasons for more males having SCD than their female counterparts cannot be immediately explained but has been linked with genetics and therefore needing further studies to help draw a valid conclusion. Moreover, there was a statistically significant association between sex and development of haemolytic crisis. This finding shows that the chance of a male child developing haemolytic crisis is increased more than twice as compared to her female counterpart. This should help clinicians and health educators educate the care givers of especially male children with SCD on the need to ensure that they provide the necessary care and make sure the haemoglobin levels of their male children are checked regularly so that where there is the need for a blood transfusion, it would be carried out quickly to prevent the development of any form of haemolytic crisis. In this present study also a greater number of the children were within the age bracket 0- 4, (46.2%) with majority of them being pre-schoolers, similar to the study by (Juwah et al., 2004). On the contrary, a study by (Brown et al., 2013) found most of the children to be in the age group 5-9 years which is also the age group with the second highest number of children in this current study. This finding shows how common SCD is among children from age 0- 9 years. Age was also found in our current study

to be statistically associated with developing pain episodes, haemolytic crisis and the development of hand-foot-syndrome. Children in the age group 1014 had a statistically significant association with developing pain episodes (P= 0.001) moreover the likelihood of developing more pain episodes increased as the child was growing up. Hence the odds of developing more pain episodes was 1.8 times for children in the age bracket 5-9 years and 4.8 times higher for children within the age bracket 10- 14 years as compared to those in the age group 0-4 years. This clearly reveals that pain episodes are more likely to increase in older (Darbari, et al, 2012) children as compared to the younger ones. It is therefore important that caregivers are educated so they understand that as their younger children with SCD grow, they will be more prone to developing more pain episodes and so the need to provide them with the necessary care in order to reduce the frequent development of pain episodes although sometimes it cannot be predicted. Again age was statistically associated with developing haemolytic crisis (P=0.037) and hand-foot-syndrome (P=0.002) specifically for children in the age group 5-9 years and 0- 4 years respectively. A child was less likely to develop hand-foot-syndrome if he was in the age group 0-4 years. Whereas a child in the age group 5 – 9 years had a 2.3 times likelihood of developing haemolytic crisis. The findings give the indication that children in the age group 0-4 are much more protected from developing hand-foot-syndrome while those in the age group 5-9 years are more prone to developing haemolytic crisis. This information should assist health educators plan educational programmes for caregivers of children with SCD in these age groups so that the occurrence of these crises can be reduced.

Findings from the current study also revealed that majority of the children live in urban areas (62.4%) with the rest living in rural communities. This finding provides information as to how possible it is to identify children with SCD in both urban and rural communities all over the country. This is in line with a study by (Jain et al., 2013) where several communities in India have children with SCD. Though there were no statistically significant association between

place of residence and the development of crises, living in an urban area lowered a child's likelihood of developing any of the crises except for splenic sequestration where living in an urban area increased the likelihood of developing it by 1.4 fold. This finding may be due to the fact that living in an urban area makes it possible to access health care readily and also provide access to a clean environment which may not be the case in most rural areas. Haemoglobin genotype SS (70.9%) was the common type presented followed by SC (22.2%) which are known to be the common types in the Ghanaian population (Ohene- Frimpong, 2008). Though there was no statistically significant association between haemoglobin genotype and the development of any of the crises in this current study, a child was 1.4 times more likely to develop pain episodes if he was haemoglobin genotype SS, 1.3 times more likely to develop haemolytic crisis and 3 times more likely to develop hand-foot-syndrome as compared to having the haemoglobin genotype SC. These findings gives the impression that children with haemoglobin genotype SS are more prone to developing crises and therefore important that clinicians and health educators provide the care givers of these children with this information and educate them to be extra vigilant so as not to expose their children to factors that may trigger crises.

5.2 Common Co-morbid Conditions presented by Children with SCD

The study also identified vaso-occlusion (44%) as the most common crisis presented by children with SCD followed by haemolytic crisis (35%), out of the 620 visits with complications to the clinic. This is similar to the study by (Teoh, et al., 2012, Brown et al., 2013).Other authors have asserted that vaso-occlusive crisis accounts for more hospital admissions in the developed world whereas infections account for admissions in developing (Fosdal and Woiner- Alexandrov, 2007) countries, this is in sharp contrast to the findings of this study where vaso-occlusive crisis was identified as the most common complication presented. Apart from vaso-occlusion which was the most common almost all the other major

crises: haemolytic crisis, hand-foot-syndrome, acute chest sequestration, splenic sequestration and then stroke were all present in children who's information were extracted, given an indication of the presence of all the major crisis in the children studied. These findings are in line with a study by (Akar and Adekile, 2008). There were also the presentation of co-morbid crisis in this study with the common one being haemolytic and vaso-occlusive crises (30%) which is in line with (Brown et al., 2013) study. This finding also makes it clear that pain episodes and haemolytic crises are very common and affects majority of children with SCD who are seen at KATH and should help in the decision making process on the kinds of interventions to put in place to reduce the frequent occurrence of these crises.

Several different types of co-morbid conditions were also identified with the common ones being infections. Malaria (20.8%) was the most common infection identified similar to the study by (Ohene-Frimpong, 2008). Other infections identified among the children were respiratory tract infection (11.6%) chronic osteomyelitis (4.9%), septicaemia (4.8%) pneumonia (3.4%), urinary tract infection (3.2%) and septic arthritis (1.9%), similar to the study by (Bansil et al., 2013). Other non-infectious complications identified included priapism, avascular necrosis of the femoral head, liver disease and parotitis. Infections are very common during the rainy seasons especially malaria, this is as a result of poor socio- economic status and unhygienic conditions under which some of these children and their families live exposing them to some of the infections mentioned above.

Findings from this study found a statistically significant association between developing pain episodes and having the malaria parasite. It was also clear that the chances of a child developing pain episodes were increased 4 times if the child had the malaria parasite. This should help clinicians and health educators provide caregivers of these children with information on how to prevent their children from being bitten by mosquitoes through the clearing of stagnant water and the provision and use of insecticide treated mosquito nets which will go a long way to

prevent them from getting malaria which is a predisposing factor for developing crises. A different study (Komda et al, 2009), however found no evidence to support the conclusion that the risk of malaria is high among children with SCD than among children without SCD which is contrary to the current study where results seem to suggest that malaria is very common among children with SCD. Having septicaemia also increased a child's chances of developing pain episodes 3.7 times. This also means that septicaemia as an infection makes the child more prone to the development of pain episodes and therefore very important for caregivers of these children to be educated on the need to as much as possible put in measures to prevent their children from getting such infections.

5.3 Proportion of Children with Stroke

Stroke, which has been identified as one of the complications children with SCD suffer from was not too common in this present study since it was present in only 1.3% of the children who took part in this study, supporting the view held by the CDC (2013), that only about 10% of children with SCD suffer from stroke complications. The findings of this study is also in line with other studies by (Jain et al, 2013; Akar and Adekile, 2013) where they identified only 0.6% and 1% of children with stroke giving an indication of an overall low prevalence of stroke in SCD children which is largely due to an increase in prophylactic red blood transfusions, routine screening of children with SCD by Transcranial Doppler ultrasonography in detecting high risk cases and other preventive measures. Most of the strokes were of the ischemic type, in line with the study by (Earley et al., 1998; Balkaran et al, 1992). Co-morbid crises children with stroke presented with included haemolytic crisis and pain episodes. There were however no statistically significant association between demographic factors and the development of stroke. However, a child was more likely, 1.3 times to develop stroke if he was in the age group 5-9 years. This findings gives the indication that children in this age group are more vulnerable to developing stroke and therefore there is the need to intensify the screening of children at this

age to make it possible to detect any signs and symptoms of stroke before it develops (Ovbiagele and Adams, 2012)

5.5 Predisposing factors for the development of complications by Children with SCD

Study findings revealed that genetic as well as environmental factors contributed to the frequent presentation of crises by children with SCD to KATH sickle cell clinic. Genetic factors such as age, and sex were found to have a relationship with the development of certain crises. Children in the age group 10 – 14 for instance had a statistically significant association with developing pain episodes (P=0.001). It was also revealed in this study that the odds of having pain episodes increased with age. This means that children in the age group 5-9 years were 1.8 times likely to develop pain episodes while those in the age bracket 10 – 14 years had a 4.8 times likelihood of developing pain episodes. Again children in the age group 0-4 years had a statistically significant association with developing hand-footsyndrome (P= 0.002) in line with the findings of the study by (Darbari et al, 2012). Whereas those in the age group 5-9 years also had a statistically significant association with developing haemolytic crisis (P=0.037). This means that children with SCD are prone to developing more pain episodes as they grow, they are also more likely to develop haemolytic crisis and therefore very important that their care givers are provided with the relevant information on how to care for them in order to ensure that the occurrence of pain episodes and haemolytic crisis are reduced as much as possible. Gender as a genetic factor was identified to have a relationship with developing haemolytic crises and being male was statistically associated with developing haemolytic crises (P=0.027). Being male puts a child at a 2.3 higher risk of developing haemolytic crises and lowers their chances of developing pain episodes, hand-foot-syndrome and splenic sequestration as compared to being female. This findings will also assist in planning educational programmes for caregivers which will help them in ensuring that they do not expose their children to factors that will make

them develop haemolytic crisis by ensuring that they are not exposed to mosquito bites which can cause malaria and may eventually lead to the development of haemolytic crisis.

Haemoglobin genotype was also identified by (Al-haggar et al, 2006, Dowling et al, 2014) to be a predictor of the severity of complications but the current study found no such association. However, a child's chances of developing pain episodes, haemolytic crisis and hand-foot-syndrome is increased 1.4, 1.3 and 3 times respectively if the child had haemoglobin genotype SS. This gives a clear indication that children with haemoglobin genotype SS are at a higher risk of developing the above mentioned complications and therefore very important that the necessary attention is given them so as to ensure that these complications are reduced to the barest minimum.

Environmental factors such as being exposed to streptococcus pneumonia, malaria and other complications like salmonella can cause serious crises (Sergeant, 2013). In this current study there was a statistically significant association between malaria and the development of pain episodes (P=0.005). This finding also makes it clear that a child is at a higher risk of developing pain episodes if he has the malaria parasite and therefore very important that clinicians and health educators educate the caregivers of these children to as much as possible provide insecticide bed nets and also keep their surroundings clear of bushes and stagnant water where mosquitoes can breed since unhygienic environments in which some of these children live exposes them to several forms of infections and other complications resulting in their frequent visits to the hospitals (Powars et al., 1991) . Even though septicaemia had no significant association with the development of pain episodes a child was 3.7 times more likely to develop pain episodes if he had septicaemia. In this study also there was no statistically significant association between septicaemia and the development of acute chest syndrome. This is however, in contrast with the study by (Fosdal and WoinerAlexandrov, 2007) where they found a statistically significant association between septicaemia and the development of acute chest

syndrome. This finding should enable caregivers protect their children from getting infections that may trigger crises and aggravate their condition.

Place of residence in this present study was identified to have no significant association with the development of crises. However a child was at a 1.4 times higher risk of developing splenic sequestration if he lived in an urban area. On the contrary a child is 0.5 times less likely to develop pain episodes, haemolytic crisis and 0.4 times less likely to develop handfoot-syndrome if he lived in an urban area. This means that if a child resides in an urban area his chances of developing splenic sequestration is higher, however living in an urban area also lowers a child's likelihood of developing pain episodes, haemolytic crisis and hand-footsyndrome. These findings are very important in the planning of interventional strategies for caregivers of children with SCD to help reduce the frequency with which these crises may occur.

Other environmental factors which have been identified by other researchers to contribute to the frequent presentation of complications included climatic conditions such as cold weather conditions and during the rainy seasons where there is an increase in the presentation of vasoocclusive crisis (Tewari and Rees, 2013).

5.6 Limitations of study

The study was only carried out at KATH and therefore findings could not be generalised to the whole SCD children population in the country. However, the results might still be widely applicable in enabling health care givers plan effective interventional strategies.

Again most of the folders extracted were incomplete, some relevant information were not available. Information on especially the socio-demographic details of care givers of children with SCD were missing and therefore made it very difficult to look at socio- demographic factors as predisposing factors to the presentation of crises which was very relevant to the study.

There were also no face to face interviews with caregivers of children with SCD as well as the children themselves, this made it impossible to get first-hand information but instead secondary data was collected which was subject to the writing of wrong diagnosis, conditions etc. which may affect the outcome of the study.

CHAPTER SIX

6.0 CONCLUSION AND RECOMMENDATIONS

6.1 Conclusion

The study identified stroke to be prevalent in about (1.3%) of the total SCD children who took part in the study. Ischemic stroke was the most common of the stroke cases. With comorbidities of haemolytic crises and vaso-occlusion among some of the children.

Majority of the children were males with most of them living in urban areas. Haemoglobin genotype SS was present in most of the children, while most of them were in the age group 04 years.

The common crises identified were vaso- occlusion, followed by haemolytic crisis and handfoot-syndrome. Infections associated with crises were malaria and septicaemia, other infections present included pneumonia, septic arthritis, urinary tract infection and respiratory tract infection. Other non- infectious complications identified were fever/ febrile illness, avascular necrosis of the femoral head and liver disease.

Predisposing factors that were associated with the development of crises included age, sex and malaria.

6.2 Recommendations

6.2.1 Education of Patients and their Caretakers

It was clear that frequent visits of children with SCD to KATH Sickle Cell clinic resulted from a number of clinical conditions, with the main ones being infections and different forms of crises. Patients and their caretakers should be educated on the causes, symptoms and complications of Sickle Cell Disease to enable them detect crises promptly and seek medical

care before they become complicated. For example, malaria was identified as a common infection that precipitate crisis. There is therefore the need for caretakers of these children to be educated on how to regularly and properly use the insecticide treated net and also clear stagnant water from their places of residence to avoid getting malaria which precipitates sickle cell crisis.

6.2.2 KATH

Hospitals like KATH which sees a lot of children with SCD complications should have a comprehensive programme that provides prompt and effective care of acute events, prophylaxis against complications, and the planning of health education programmes that will help these children and their care givers recognise complications and seek prompt medical care.

Clinicians, nurses and other hospital staff who are involved with the care of children with SCD should be equipped with the current and relevant information on how to promptly detect and treat complications before they become severe. Health educators of these children and their care givers should be well- versed with the causes, symptoms, complications as well as the predisposing factors to the development of crises so that they can properly educate caregivers of these children to ensure that they provide the proper care for their children in order to reduce the occurrence of crises and other complications

6.2.3 MOH

The Ministry of Health should formulate policies that will ensure the compulsory screening of babies at all health facilities to ensure the early detection of SCD so that these babies can be enrolled into care at the earliest possible time and then given a comprehensive care which will involve the prompt treatment of complications and the education of care givers on the

predisposing factors to the development of complications. This will ensure early detection of crises and complications and therefore assist caregivers in seeking medical care for their SCD children at the earliest possible time.

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APPENDIX A

ABSTRACTION FORM FOR COLLECTION OF DATA ON CHILDREN WITH SCD AT KATH

SECTION 1. DEMOGRAPHIC DETAILS OF PATIENT

PATIENT ID ……..

1. Age ……..

2. Sex

(a) Male

(b) Female

3. Place of Residence …….

4. Educational Background of patient

(a) Nil

(b) Crèche

(c) KG

(d) Nursery

(e) Primary

(f) JHS

5. Religion

(a) Muslim

(b) Christian

(c) Atheist

6. Educational Background of Caretaker of patient. (a) Nil

(b) Primary

(b) JHS

(d) MSLC
(e) SHS

(f) Tertiary

7. Occupation of Caretaker of patient……

8. Does patient have siblings suffering from SCD?

(a) YES
(b) NO

9. If Yes to (8) how many ……

SECTION 2. INITIAL ASSESSMENT DETAILS OF PATIENT

10. Date Diagnosed with SCD ……..

11. Haemoglobin Phenotype ……

12. Date of first visit to Sickle Cell Clinic……..

13. Haemoglobin level at initial visit……..

14. Type of crisis presented at initial visit.

( ) None
( ) Hand –foot-Syndrome
( ) Pain episodes
( ) Anaemia
( ) Acute Chest Syndrome
( ) Splenic Sequestration
( ) stroke
Others specify……

15. Associated infections and other complications ( ) None
( ) Septicaemia
( ) Malaria
( ) Pneumonia
( ) Meningitis

( ) U T I
( ) Arthritis
( ) Pharyngitis

Other specify …….

SECTION 3. FOLLOW-UP DETAILS OF PATIENT

Patient ID ……..

Follow-up visit Number ……..

16. Date of follow-up visit…….....

(I) Type of visit

(a) Schedule visit

(b) Sick Visit

(II) Type of crisis presented.

( ) None

( ) Hand –foot Syndrome

( ) Pain episodes

( ) Anaemia

( ) Acute Chest Syndrome

( ) Splenic Sequestration

( ) stroke

Others specify……

17. Associated infections and other complications

( ) None

( ) Septicaemia

( ) Malaria

( ) Pneumonia

( ) Meningitis

( ) Urinary Tract Infection

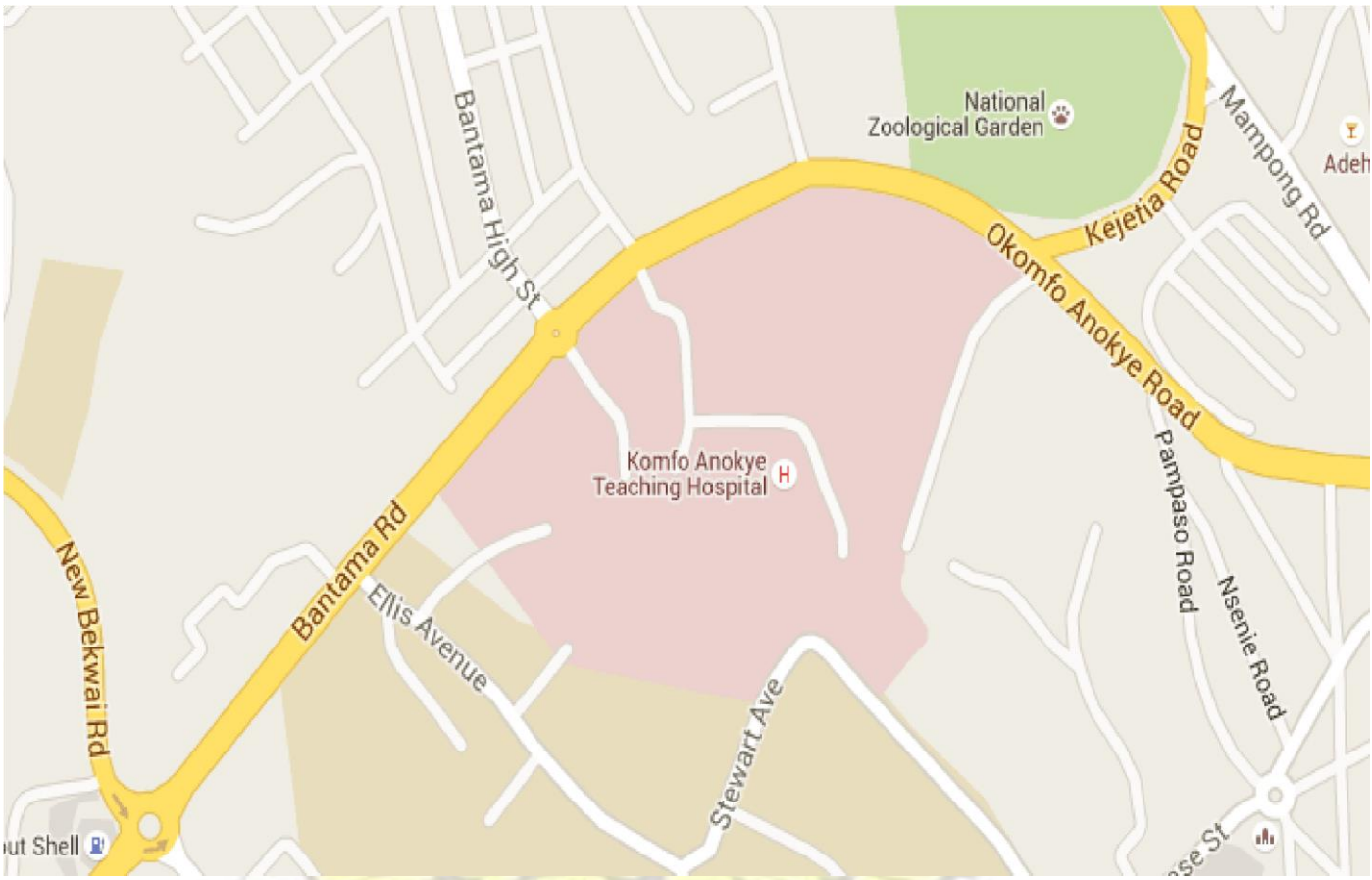
( ) Arthritis

( ) pharyngitis

Other specify……

(iii) Haemoglobin level ……

18. Total number of visits to Sickle Cell Clinic…….



APPENDIX B

Komfo Anokye Teaching Hospital on the map