DIETARY INTAKE AND NUTRITIONAL STATUS OF CHILDREN AGED 3-12 YEARS WITH SICKLE CELL DISEASE

BY

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JULY, 2016
DECLARATION

I, Isaac Boadu, hereby declare that this work is the result of my own research and that this dissertation has neither in whole or in part been submitted to this University or elsewhere for another degree. All references to other people’s work which served as a source of information in this research have been duly acknowledged.

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DEDICATION

I dedicate this work to my younger sister, Rebecca Boadu, and all families with children who have sickle cell disease. Let us have time and patience for these innocent children.
AKNOWLEDGEMENT

I would like to thank God for how far he has brought me. I never imagined. All glory and honour be unto your Holy name.

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To all ‘I Say God Richly Bless You’.
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ABSTRACT

Background: Sickle cell disease (SCD) is a chronic genetic blood disorder common among people of African descent. The disease places nutritional burden among affected individuals which affect their nutritional status. The aim of the study was to determine the dietary intake and nutritional status of children with sickle cell disease and determine the associations of severity of illness with dietary intake and nutritional status.

Methodology: A cross-sectional study was carried out in 120 children with SCD aged 3-12 years at the paediatric Outpatient Department of Princess Marie Louise Hospital (PML) in Accra. A semi-structured questionnaire was used to take information on participant’s demographic characteristics, and clinical data were obtained from their medical records. Height and weight were measured for participants and their haemoglobin levels were determined to assess their anaemia status. Dietary data was obtained using a single 24-hour dietary recall and food frequency questionnaire.

Results: Nutrient intakes were low particularly for calcium and antioxidant nutrients (vitamin C and E) and this declined with increase in age. Malnutrition was recorded for 38% of the children with prevalence of stunting, underweight, thinness and wasting as 25.8%, 20.0%, 15.8% and 6.8% respectively. Having sickle cell anaemia (SS) was significantly associated with stunting (OR 4.8, 95%CI, 1.4-16.1) but not underweight. Almost all the children (98.3%) were anaemic. There was no relationship between dietary intake, severity of illness and nutritional status among the study children.

Conclusion: Nutrient intakes were generally below recommendations among the participants. There is the need to develop comprehensive management coupling nutritional therapy to medical care to improve the lives of children with SCD. Nutritional management should focus much on calcium-rich foods and antioxidants nutrients particularly vitamin C and E to reduce rapid erythrocyte haemolysis and chronic anaemia.
CHAPTER ONE

1.0 Introduction

1.1 Background

Ghana like many other African countries faces the double burden of infectious and chronic diseases including sickle cell disease (SCD), the underappreciated and least addressed contributor to global childhood morbidity and mortality (Daak et al., 2016). SCD is recognised as one of the common chronic genetic disorders in humans with variable clinical manifestations. The disease is characterized by the predominance of sickle haemoglobin (HbS) produced as a result of a point mutation where a non-polar amino acid, valine, substitutes a polar amino acid, glutamic acid, in the β-globin chain of haemoglobin molecule (Moheeb et al., 2007). The main consequences resulting from this abnormality are vaso-occlusive events and increased haemolysis. Vaso-occlusive events may lead to tissue, bone, and organ damage while chronic haemolysis may lead to anaemia with a base line haemoglobin level as low as 6.0g/dl (Stuart and Nagel, 2004). Comorbidities such as pain, stroke, anorexia associated with SCD have been shown to decrease dietary intake which results in impaired growth, poor nutritional status and delayed skeletal and sexual maturation (Zemel et al., 2007). These clinical features are common in children with type SS, the most severe form of SCD (Esezobor et al., 2016). Although the exact reasons for the poor growth have not been elucidated, some suggested reasons include hypoxia caused by reduced red blood cell levels and associated increase in energy and nutrient requirements, adverse effects of vaso-occlusion, endocrine dysfunction, and chronic organ damage (Al-Saqladi et al., 2008).

At birth, infants with SCD-SS are normal in size due to predominant foetal haemoglobin, but significant growth impairment become apparent by 5 years of age (Zemel et al., 2002). Majority of children born with SCD occur in the developing world, with an estimated 200,000 annual HbSS births in sub-Saharan Africa (Ansong et al., 2013). In Ghana, recent studies indicate that 2% (14,000) of annual newborns are affected by SCD and 25% of the Ghanaian population are
carriers of the sickle cell gene. One in three Ghanaians have the haemoglobin S and/or C gene (Edwin et al., 2011). Despite the high mortality rate associated with SCD, the disease is not listed as a major contributor to mortality in children less than 5 years of age, even in countries where 1%-2% of all births may be affected by SCD. This is most commonly due to the lack of awareness of the underlying SCD diagnosis and consequences on childhood mortality. Undoubtedly, one can argue that children with SCD are underrepresented within the top 5 global leading causes of death (Pneumonia, diarrhoea, meningitis, malaria, and sepsis) of children under 5 (Liu et al., 2015) since SCD children are all susceptible to these diseases, but without pre-existing diagnosis these conditions may simply be diagnosed as pneumonia, sepsis, etc.

Information on SCD has mainly been reported among US population, India and Jamaica with less information in Africa where the gene is highly predominant. Although some studies have reported on nutritional status of children with SCD in some parts of Africa (Cox et al., 2011, Tsang et al. 2014), there has been paucity of studies designed to identify the dietary intake and nutritional status of children with SCD in Ghana where prevalence is 2% and newborn screening is yet to be scaled up nationally. A study in Ghana reported high prevalence of malnutrition (61.3%) among children with SCD but failed to consider their dietary intake (Osei-Yeboah et al., 2011). Emphasis on dietary intake and its association on nutritional status and disease severity among children with SCD have thus, not been adequately documented in Ghana. The purpose of this study was to assess the dietary intake and nutritional status of children aged (3-12 years) with SCD.
1.1 Rationale

SCD contributes to 9-16% of infant mortality in Ghana annually (Sickle Cell Foundation Ghana, 2015). Nutritional status of children with SCD cannot be overlooked when considering the health of a nation, especially developing countries such as Ghana where the prevalence of SCD has been reported as 2% while 25% of the population are carriers of the sickle cell gene. Approximately half of these children (SCD) die in the first five years if they are undiagnosed and untreated (Sickle Cell Foundation Ghana, 2015). Currently, there is no nationwide standardised comprehensive management of SCD including dietary recommendations among children in Ghana. This is partly due to a dearth of information on the dietary intake and nutritional status which may be a contributing factor to the increased complications, morbidity and mortality among children with SCD. It is therefore necessary to consider the health of these patients in-terms of dietary intakes to help develop a comprehensive national management plan in the domain of nutritional regimen and medical care for people with SCD.

1.2 Research questions

- What is the current nutritional status of children with sickle cell disease?
- What are the dietary intakes of children with sickle cell disease?
- Is there a relationship between severity of illness and nutritional status?

1.3 Main objective

The main objective of this study was to assess the dietary intake and nutritional status of children with sickle cell disease and its association with disease severity.
1.3.1 Specific Objectives

- To assess the dietary intake of children with SCD
- To determine the anthropometric indices of children with SCD
- To estimate the prevalence of anaemia among children with SCD
- To assess the relationship between dietary intake and disease severity among study participants.
- To determine the relationship between nutritional status and disease severity.

1.4 Hypotheses

- Children with sickle cell disease have reduced dietary intake and poor nutritional status.
- Severity of sickle cell disease is associated with poor dietary intake and nutritional status.
CHAPTER TWO

2.0 Literature review

2.1 History and meaning of sickle cell disease (SCD)

Even though sickle cell disease was speculated to exist among one Ghanaian family as early as 1670 (Abramson et al., 1973), the disease was first described and reported in a Southern Journal of Medical Pharmacology in 1846, USA, where a runaway slave charged with murder was described in a paper titled “case of absence of the spleen”. After autopsy, the physician noted unusual body builds and described it as a man who lived without the spleen. It was later reported in African medical literature in 1870s, when it was known locally as ogbanjes (“children who come and go”) because of the high infant mortality rate associated with the condition (Ebrahim et al. 2010). In 1910, Ernest Edward Irons, an intern of James B. Herrick found "peculiar elongated and sickle-shaped" cells in the blood of Walter Clement Noel, a dental student from Grenada who was admitted several times at Chicago Presbyterian hospital following anaemia diagnosis (Herrick, 2000).

In 1922, the disease was named "sickle-cell anaemia" by Vernon Mason and in 1949, Pauling and colleagues first demonstrated that SCD occurs as a result of an abnormality in the haemoglobin molecule and described it as a ‘Molecular Disease’ (Rees et al., 2010). Since its discovery, the focus on the metabolic pathways and pathophysiology of the disease manifestations have increased resulting in effective management and treatment, and reduced morbidity and mortality as well as increased life expectancy among patients in some parts of the world (Rees et al., 2010).

SCD comprise a group of genetic abnormalities that affects the synthesis of haemoglobin.

The disease is characterized by a replacement of a non-polar amino acid, valine, for a polar amino acid, glutamic acid, in the β-globin chain of haemoglobin molecule (Mir et al., 2016). SCD mostly affects people of African or Hispanic descent with the majority of cases in sub-
Saharan Africa. The consequence of this point mutation is the formation of haemoglobin S (HbS).

The presence of the non-polar amino acid at the β-globin chain stimulates polymerisation of haemoglobin under low oxygen conditions, which changes the normal biconcave shape of red blood cells into a sickle shape and decreases their elasticity (Platt and Sacerdote, 2006). When sickle haemoglobin is deoxygenated, it forms large insoluble polymers. This distortion changes the rheological properties of red blood cells (RBCs) and blocks blood flow in small peripheral vessels. Major consequences include vaso-occlusion; haemolysis marked by anaemia and increased susceptibility to infections (Brunno and Gianni, 2016). People who inherit a copy of the abnormal gene are described as carriers whilst those who inherit the sickle-cell genes from both parents develop SCD. People who are carriers are often asymptomatic but are capable of transferring the gene on to their children.

2.2 Sickle cell epidemiology

Sickle cell disease affects almost all populations across the globe with majority (70%) of the disease occurring in Africa (Justus et al., 2015). About 300,000 annual births of children with SCD occur in the world. In 2010, two-third of SCD births was documented in sub-Saharan Africa making it the most burdened region (Piel et al., 2013). There is an increased prevalence among the African as well as the Asian populations. The sickle cell gene also has a wide spread distribution in some parts of America, Europe and the Caribbean. This is mainly due to the Atlantic slave trade as well as the economic migration of Africans to different parts of the world. In Germany and UK for example, there are currently an estimated 1000 and 12,500 people with SCD respectively (Jaeckel et al., 2010). In United States, SCD affects approximately 100,000 people and each year 2000 new diagnoses are detected via newborn screening (Arnold et al., 2015). In India, the prevalence of beta sickle (βS) gene varies from 0-40% in different population group (Mukherjee and Gangakhedkar, 2004). In sub-Saharan African countries such as Cameroon, Republic of Congo, Gabon, Ghana and Nigeria, the prevalence of sickle cell trait
and SCD has been estimated to be 25% and 2% respectively. Nigeria has been reported to have the highest prevalence of SCD in the world (Adegoke and Kuteyi, 2012). In countries where the trait prevalence exceeds 20%, the disease affects about 2% of the population (WHO, 2010). For example in Ghana where the sickle cell trait is 25%, it has been documented that about 2% of neonates are affected by SCD leading to 14,000 new cases annually (Wilson et al., 2012).

Sickle cell trait is known to show a protective effect against malaria, and this may be the reason why it is prevalent among the Africa populace (Bartolucci, 2014).

2.3 Haemoglobin molecule and pathophysiology of SCD

Haemoglobin (Hb) is a polypeptide tetramer composed of α-like and β-like globin subunits, each bound to a haem prosthetic group (Bhasin and Walter, 2007). The major functions of haemoglobin are to transport oxygen (O₂) from the lungs to peripheral tissues and carbon dioxide (CO₂) from the tissues to the lungs. The globin polypeptides are synthesised from separate α-like and β-like globin genes located on human chromosomes 16 and 11 respectively (Steensma et al., 2005). Normal adult humans have haemoglobin A, which consists of four subunits, two alpha and two beta (α2β2) chains, haemoglobin A2, which consists of two alpha and two delta (α2δ2) chains and foetal haemoglobin (Hb F), consisting of two alpha and two gamma chains (α2γ2) (Bhasin and Walter, 2007). The predominant haemoglobin in foetal life and early infancy is Hb F which declines approximately 6 months after birth and is gradually replaced by adult haemoglobin (Hb A) (Waye and Chui, 2001). Haemoglobinopathies are chronic inherited disorders that occur due to defects in the synthesis of α and/or β-globin chains of haemoglobin molecule. The disorder can result in the production of structural defects of the haemoglobin (Hb) molecule or quantitative defects in one of the globin chain (α or β) (Platt and Sacerdote, 2006). Structural Hb variants are mainly due to a single amino acid substitution (point mutation) in one of the globin chains which does not only distort the secondary and tertiary structure but also the morphology of Hb molecule. This subsequently affects the rheological and functional properties (Joiner, 1993). The red blood cells assume the shape of a ‘sickle’ under low oxygen conditions.
with vaso-occlusion and chronic haemolysis being the main consequences of this disorder (Clarke and Higgins, 2000). Healthy red blood cells have on average 120 days lifespan but sickle cells survive for only 10-20 days. Although the bone marrow tries to compensate for the rapid loss of red blood cells it does not balance the rate of destruction resulting in anaemia (Tarer et al., 2006).

2.4 Types of Sickle cell disease

There are different types of SCD depending on the type of haemoglobin gene inherited from parents. SCD is described within four major genotypes: homozygous sickle cell (SS) disease, sickle haemoglobin C (SC) disease, sickle cell β+ thalassemia (Sβ+ thalassemia) and sickle cell β0 thalassemia (Sβ0 thalassemia) (Al-Saqladi et al., 2008). These different genotypes are associated with adverse health outcomes with varying severity and potential effect on nutritional status.

The most common and clinically severe form of SCD is haemoglobin SS often referred to as sickle cell anaemia and Sβ0 thalassemia (Kanter and Kruse-Jarres, 2013). People suffering from sickle cell anaemia (SS) experience severe complications such as pain, serious infections, chronic anaemia and organ failure compared to the other heterogeneous types of the disease (HbSC, HbS/β0, HbS/β+) (Hyacinth et al., 2010). In Ghana, the common genotypes identified are ‘SS’, ‘SC’, ‘SD’ and Sβ thalassemia (Osei-Yeboah and Rodrigues, 2011).

2.4.1 Sickle cell anaemia (SS)

Haemoglobin (S) was among the first type of haemoglobin variants characterised and identified (Allison, 1965). The homozygous haemoglobin (SS) is often referred to as ‘sickle cell anaemia’, the most common type of SCD. It occurs when copies of abnormal haemoglobin S gene is inherited from both parents. In Hb S, hydrophobic valine is substituted for the normal hydrophilic glutamic acid at the 6th residue of the beta globin gene due to single nucleotide mutations (GAG/GTG) (Hoban et al., 2015). When Hb S is deoxygenated, it tends to aggregate and polymerizes which then assume the abnormal "sickle" shape. The rate and degree of
polymerization determines the rheological properties of the sickle erythrocytes. The consequences of the polymerisation results in severe complications including vaso-occlusive events (bone pain, tissue and organ damage) and increased haemolysis leading to anaemia (Stuart and Nagel, 2004). Other pathological conditions common among sickle cell anaemia patients include ocular manifestations, respiratory diseases of the lower and upper airways, pulmonary hypertension, reduced dietary intake, poor growth and delayed maturation (Zemel et al., 2007).

2.4.2 Sickle haemoglobin C (SC)

Heterozygous sickle haemoglobin (SC) is the second most frequent haemoglobinopathy after sickle cell anaemia and is prevalent among many West African countries (Weatherall, 2010). It results from another point mutation of lysine for glutamic acid GAG/AAG at the 6th codon of the β-globin gene (Modiano et al., 2001). Hb C promotes the stimulation of K:Cl co-transport and this leads to dehydration and deformed red blood cells (Joiner, 1993). Patients with sickle haemoglobin C (SC) have less clinical consequences (mild haemolytic anaemia and decreased pain episodes compared to homozygous haemoglobin S (Hb SS) (Markham et al., 2003). However, an increase in blood viscosity is common among this genotype and is thought to increase complications, such as retinopathy and renal papillary necrosis (Markham et al., 2003).

2.4.3 Thalassemia

Unlike a single amino acid substitution that result in abnormal form of haemoglobin C and S, thalassemias (α and β) occur due to quantitative reduction in the synthesis of the globin chain of haemoglobin molecule (Allen et al., 1997). Their classification is based on the specific globin chain that is ineffectively produced. Disorders with reduced β-globin chains are termed β-thalassemia whereas those with decreased α-chain production are referred to as α-thalassemia. The extent of severity and clinical manifestations of a particular thalassemia depends on the amount of globin chain produced and the stability of the residual excess chains (Siriratmanawong et al., 2001).
2.5 Sickle Cell Trait

People who are heterozygotes for HbS or HbC are said to have sickle cell trait (SCT), rather than SCD and they are generally asymptomatic (Eckart et al., 2004). It is characterised by the inheritance of a normal beta haemoglobin gene (HbA) from one parent and a sickle cell gene, often haemoglobin S (HbS) from the other parent. In this condition, every red blood cell contains both haemoglobins A and S, with haemoglobin A being the predominant haemoglobin as opposed to SCD in which sickle haemoglobin (S) is the predominant haemoglobin (Jordan et al., 2011).

The prevalence of SCD depends on sickle cell trait. Where the prevalence of SCT exceeds 20%, SCD is estimated to be at least 2% (WHO, 2010). There is high prevalence of SCT (25%) in Sub-Sahara Africa and this has been attributed to the protective effect of HbS against both severe and uncomplicated malaria (Kreuels et al., 2009). Although the protective role is established, the mechanism behind has not been well elucidated. Possible mechanisms includes; parasite growth inhibition due to low oxygen, specific intra-erythrocytic conditions of HbAS red blood cells, such as low intracellular potassium, and reduced parasite growth due to translocation of host micro-RNA to the parasite (Modiano et al., 2001). Other mechanisms include impairment of rosette formation due to decreased oxygenation, decreased cyto-adherence and rapid degradation of sickled red blood cells making it difficult for complete cycle of the malaria parasites (Gong et al., 2013). The survival advantage of Hb AS carriers in malaria endemic areas may be attributed to the lower risk of chronic malnutrition in early childhood, mediated by protection against mild malaria episodes (Kreuels et al., 2009).

2.6 Sickle Cell Diagnosis

The diagnosis of SCD is based on analysis of haemoglobin. In most laboratories, sodium metabisulphite is applied to a drop of blood, which reduces the oxygen tension inducing the typical sickle-shaped red blood cells seen under the microscope. If the screening test is positive,
further test such as haemoglobin electrophoresis, isoelectric focusing, and high performance liquid chromatography (HPLC) is done to determine the genotype (Rees et al., 2010). Although new-born screening and early diagnosis is important to give children timely and better treatment, this is not a common practise in most African countries. Diagnosis is usually done when severe complications start occurring later in life.

2.7 Complications of sickle cell disease

Although SCD is a disease that affect red blood cells, it virtually affects every organ in the body due to decreased oxygen supply (Costa and Fertrin, 2016). The clinical consequences can be divided into 4 groups: haemolysis, vaso-occlusion, infection, and organ dysfunction (Hyacinth et al., 2010). In a study by Olabode and Shokunbi (2005) to determine the types of crises presenting in sickle cell patients, it was found out that vaso-occlusive, sequestration, infarction, aplastic and haemolytic were the major types of crises occurring in the study participants.

2.7.1 Vaso-Occlusion

Sickle vaso-occlusion is the process by which normal tissue perfusion is interrupted by sickle erythrocytes (Steinberg, 2008). This is the most common type of complication and cause of hospitalization occurring among sickle cell patients particularly those homozygous for SCD (SS). Sickle red blood cells may block circulation, particularly in the post-capillary venule and this reduces blood flow to vital organs leading to hypoxia, ischemia, necrosis, painful episodes, acute splenic sequestration, and priapism (painful and prolonged penile erection). Affected sites normally include the arms, legs, back, abdomen, chest, and head. Vaso-occlusion crisis is often associated with poor quality of life and increased risk of death among affected individuals (Almeida and Roberts, 2005).

2.7.2 Haemolysis and Anaemia

Haemolysis occurs when there is damage to red blood cell membrane which leads to the cells breakdown resulting in anaemia. The spongy bone marrow is responsible for the production of red blood cells to replace old ones (Almeida and Roberts, 2005). In a normal individual, red
blood cells have an average lifespan of 120 days unlike 10-20 days in sickle cell anaemia. Although the bone marrow attempts to compensate for the rapid degradation of the red blood cells, it does not match the constant loss leading to low haemoglobin concentrations (anaemia). Haemolysis is also associated with narrowing or increased pressure in blood vessels and may lead to complications such as pulmonary hypertension and skin ulcers. Studies have respectively recorded a lower baseline haemoglobin (8.6g/dl, 6.7g/dl, 7.5g/dl) among sickle cell patients compared to healthy controls (14g/dl, p<0.00001, 7.6g/dl, p<0.0001, 11.0g/dl, p<0.001) (Buchowski et al., 2002; Daak et al., 2013; Cox et al., 2011). This is confirmed by a recent study by Nikhar et al., (2012) who recorded a lower mean haemoglobin level among sickle cell patients from rural and urban areas in India with a mean haemoglobin of 7.8 ± 0.2 g/dl and 9.1 ± 0.4 g/dl respectively.

2.7.3 Infection

Frequent infection especially from bacteria predominantly *Streptococcus pneumoniae*, is a major problem and the primary cause of death particularly in children with SCD (Helvaci, 2016). This is as a result of immune function impairment, decreased dietary intake and early loss of splenic function. Before the introduction of prophylactic penicillin administration, children with SCD were more prone to developing pneumococcal disease, manifesting as septicemia or meningitis (Halasa et al., 2007). Individuals affected with SCD are susceptible to infection, especially to bacterial pathogens which promote pathophysiological changes such as acute chest syndrome, painful episodes and hyper haemolytic episodes (Ansong et al., 2013). Although this has not being clearly understood, it is believed that malnutrition from increased demand of certain nutrients may be the cause (Miller et al. 2000). The interaction of nutrition and pathophysiology of SCD is conceptually presented below:
2.7.4 Organ dysfunction
End organ damage results from reduced oxygen circulation to vital tissues. Major organs affected include the brain, lungs, kidney, eyes, genitals and the spleen (Kanter and Kruse-Jarres, 2013). In some instances some of these organs become so severely damaged that they may require surgical attention to keep the patient alive.

2.8 Severity of sickle cell disease
SCD is associated with adverse health outcomes with varying severity which affects nutritional status. Under-nutrition is associated with poor clinical outcomes and severity of disease in children with SCD. The main cause of disease severity is the rate and degree of HbS polymerisation coupled with other environmental factors (infections, nutrition and socio-economic status). These may influence disease complications and the rate of survival (Makani et al., 2007). The manifestations and markers of severity determinants can be number of hospitalization, blood transfusion and lower haematological indices (Zemel et al., 2007).
In Jamaican children, levels of haemoglobin (Hb) decreased (8.4g/dl to 7.6g/dl, p=0.03) as well as foetal haemoglobin (Hb F) (8% to 3.8%, p>0.0001) with increasing number of hospitalisation (Singhal et al. 1996). A recent study has showed that inadequate nutritional intake has a significant impact on SCD severity indices (Mandese et al., 2016).

2.9 Management of sickle cell disease

Like many chronic diseases, SCD cannot be cured. However, the disease can be managed such that disease severity and complications are minimized by high fluid intake, healthy diet, folic acid supplementation, pain medications, antibiotics for infections, and specific treatments such as hydroxyurea (Ebrahim et al., 2010).

SCD patients have special (increased) needs and require a comprehensive care which may involve a multi-disciplinary team of well trained professionals (medical and non-medical services) and understanding social support system that meets the physical, emotional, psychological as well as the financial needs of patients with SCD (Edwin et al., 2011). In the last 50 years, survival has improved dramatically for people with SCD due to strategies put in place in the management of the disease. Sickle cell patient’s average life expectancy in the 1970s was 20 years of age (Platt et al., 1994). By the early 1990s, the Cooperative Study of SCD estimated a median life expectancy of those with sickle cell anaemia, the most severe form of the disease, to be 42 years of age for males and 48 years of age for females (Platt et al., 1994). In developed countries like Great Britain and USA, treatment methods include bone marrow transplant the only curative care, provision of hydroxyurea, prophylactic penicillin, blood transfusion and early detection measures such as new-born screening (Ansong et al., 2013). Although impressive measures such as new-born screening, blood transfusion, provision of analgesics are put in place in the management of sickle cell patients, the cost effective and potent drug, hydroxyurea which has been shown to decrease the number of days of hospitalization and transfusion, lower crises rate, and lessen acute chest syndrome (Chawla et al., 2013) is yet to be used in most developing countries. In a recent study by Nsiah and colleagues (2014) to determine the drug regimen
prescribed for sickle cell patients attending a clinic in Kumasi, Ghana, it was concluded that the drugs prescribed were routine drugs including analgesics, anti-malarials, antibiotics and haematinics. The top ten commonly prescribed drugs were folic acid, diclofenac, ibuprofen, B-complex, atesunate/amodiaquin, paracetamol, penicillin V, amoxiclav and zincovit.

2.9.1 Hydroxyurea

Hydroxyurea (HU), an antimetabolite, was the first drug approved for the effective management of SCD (Braga et al., 2005). Studies (Fersta et al., 2011, Steingberg et al., 2003) have reported on the beneficial effect of the drug including decrease in the number and frequency of hospitalisation, decrease rate of transfusion, reduce painful episodes (crises) and improve the quality of lives of patients with SCD. Although the known efficacy and safety of hydroxyurea for the treatment of SCA is approaching 30 years (McGann et al., 2016) the drug is yet to be used routinely in countries that are not medically or economically advanced including Ghana, where the need is undoubtedly the greatest. The main clinical benefit of HU is derived from its capacity to increase foetal haemoglobin (HbF) production as well as decrease production of leukocytes and reticulocytes that may contribute to vaso-occlusion. This subsequently decrease the rate of polymerisation of sickle haemoglobin (S) (Wali and Moheeb, 2011).

2.9.2 Folic acid and penicillin

Management of SCD patients start with early diagnosis, preferably in the new-born period and include penicillin prophylaxis, vaccination against pneumococcus bacteria, and folic acid supplementation. Folic acid is widely prescribed for SCD patients possibly due to its role in the synthesis of new red blood cells to compensate for the rapid degradation of red blood cells which often results in anaemia. Although there is continuing debate about the efficacy of folic acid supplements in the management of SCD, particularly reports on masking vitamin B12 deficiency, it has been reported to improve growth, reduce dactylitis and reduction of homocysteine levels leading to reduced cardiovascular, stroke and venous thrombosis risk (Al-Yassin et al., 2012). Other investigators also found no significant differences (p<0.0001) in
haemoglobin, growth characteristics, acute splenic sequestration, dactylitis, episodes of bone or abdominal pain in a double blind controlled trial of folic acid supplementation in homozygous sickle (SS) children over one year period (Rabb et al., 1983). In Ghana, as part of medical care, 5mg/d of folic acid supplement is routinely prescribed for SCD patients (Nsiah et al., 2014).

2.10 Nutritional care and intervention

Nutrition is an important but often forgotten aspect of care of patients with SCD despite the well-established facts on the benefits of nutrient (s) supplementation on growth and nutritional status. These patients often report decreased appetite, possibly because of the chronic inflammatory state and inadequate education concerning nutritional requirements which results in several nutritional deficiencies and poor clinical outcomes. Nutrient deficiencies are associated with immunologic, growth and maturation abnormalities (Hyacinth et al., 2013). Although nutrient deficiencies that occur in SCD are poorly understood, several factors have been proposed for the limited energy and nutrients observed among these patients. These include; reduced intake potentially from the anorexic effects of comorbidities such as pain and stroke, decreased absorption of nutrients, increased degradation and losses of nutrients, increased requirements as a result of elevated basal metabolic rate and alterations in metabolic pathways (Cox et al., 2011).

2.10.1 Macronutrients

Patients with sickle cell anaemia have greater than average requirements for both protein and energy when compared with their sex and age matched peers (Salman et al., 1996). This increased requirement is poorly understood but it has been suggested that hypermetabolism due to reduced life-span of erythrocytes places an increased demand on protein stores, increases protein turnover and consequently increased energy expenditure (Salman et al., 1996).
2.10.1.1 Protein and energy

Protein is an essential macronutrient needed to promote growth in children and synthesise haemoglobin. Children with SCD require adequate amounts of protein and energy (more than RDA) to maintain normal metabolism and physiological processes (Hyacinth et al., 2010). Increased protein turnover have been reported among SCD patients and this adds an additional nutritional burden (Borel et al., 1998). Heyman et al (1985) were the first to report on insufficient macronutrient intake in a supplementation trial. The researchers studied five malnourished children with HbSS and reported that two of the children showed improvement in growth after naso-gastric supplements of protein and calories. Although a firm inference of their report is limited due to the small sample size, the results shows the beneficial outcome of protein supplements in the management of malnutrition in SCD patients. Similar evidence has been observed in animal studies where the researchers investigated the efficacy of increased dietary protein for improving weight gain and reducing inflammation in Berkeley sickle cell mouse (Archer et al. 2008). Currently, there are no special dietary recommendations for protein and/or energy for patients with SCD despite the sufficient information to suggest that increased dietary requirements as much as needed in pregnancy and growth is necessary to improve clinical outcome of the disease (Hyacinth et al., 2010).

2.10.1.2 Fatty acids

Fatty acids are important components of cell membranes and are precursors to many other substances in the body. They may play important role in improving and maintaining the overall health and clinical outcomes in patients’ with SCD. Fatty acids especially essential fatty acids (Omega-3 and Omega-6) are needed to synthesise and repair cell membranes, promote growth of children particularly for neural development and maturation of sensory systems.

Omega-3 (n–3) fatty acids (DHA and EPA) have been shown to have anti-aggregatory, anti-adhesive, anti-inflammatory, and vasodilator properties (Daak et al., 2013). Studies have reported on the beneficial effect of fatty acids on the clinical outcomes of sickle cell patients.
particular in reduction of painful episodes (Tomer et al., 2001). In a randomised double bind placebo controlled trial to determine the effect of omega-3 (n−3) fatty acid supplementation in patients with sickle cell anaemia in Sudan, Omega-3 treatment reduced the rate of clinical vaso-occlusive events from at least 1 to 0.21 (95% CI: 0.09, 0.47; P < 0.001), severe anaemia (3.2% compared with 16.4%; P < 0.05) and blood transfusion (4.5% compared with 16.4%; P < 0.05) (Daak et al., 2013). Butyrate, a short chain fatty acid have also been reported to stimulate the production of foetal haemoglobin, which decrease polymerisation of sickled erythrocytes (Liakopoulou et al., 1995). In animal studies, dietary supplementation of docosahexanoic acid (DHA) increases red blood cell membrane flexibility and reduces irreversibly sickled cells by 40% in SS mice (Wandersee et al., 2015). These results point to the potential therapeutic benefits of dietary omega-3 fatty acids in SCD.

2.10.2 Micronutrients

Micronutrients are essential vitamins and minerals required in small quantities for good health. Their deficiencies have been associated with sickle cell disease. These include iron, zinc, copper, folic acid, pyridoxine and vitamin E (Hyacinth et al., 2010). Possible mechanism by which micronutrient deficiency may develop in sickle cell disease include decreased intake, intestinal malabsorption, increased urinary loss and catabolism of specific nutrients (Hyacinth et al., 2010).

2.10.2.1 Iron

Iron is an important trace mineral required for haemoglobin synthesis and proper immune function. Good sources of iron include meat, poultry, whole grains, liver, beans and eggs. Although iron is an important component of red blood cells, excess of it may contribute to generation of free radicals to cause oxidative stress and damage red blood cell membranes. This may consequently lead to increased haemolysis in patients with SCD (Pack-Mabien et al., 2015). It has been suggested that diet for sickle cell patients should be low in absorbable iron but high in vegetable proteins. Thus, iron-rich foods, such as liver, iron-fortified formula, iron-
fortified cereals, and iron-fortified energy bars should be excluded (Mahan et al., 2012). Deficiency of iron results in anaemia and poor immune function with symptoms manifesting as body weakness.

There are discrepancies in studies from developing and developed countries regarding amount of iron stores in sickle cell patients. While studies from most developing countries have found low iron stores in their sickle cell participants (Vichinsky et al., 1981, Okeahialam and Obi 1982), the vice versa occurred in the developed world (Harmatz et al., 2000). This could be explained partly by the lower socioeconomic status among the developing world which affects dietary intake of iron and/or chronic transfusion therapy.

Although iron deficiency anaemia is a problem in many developing countries, it is often uncommon among patients with SCD because of increased gastrointestinal absorption associated with haemolysis and the iron provided by red cell from blood transfusions (Vichinsky et al., 1981). However, it is worthy of note that iron deficiency may be present in some patients with SCA owing to repeated phlebotomies and haematuria secondary to renal papillary necrosis. This should be assessed, and the diet adjusted accordingly (Mahan et al., 2012).

2.10.1.2 Zinc

Zinc is the second most abundant trace element in the body next to iron (Mahan et al, 2012). It is involved in all major aspects of cellular functions including metabolism, detoxification, antioxidant defences, signal transduction, and gene regulation (Bao et al., 2008). Consequences of zinc deficiency include anorexia, growth retardation, neurosensory defects, and immune dysfunction in humans (Prasad, 2008).

In the context of SCD, more attention has been focused on zinc than any other mineral. The deficiency of zinc has been reported more among people with ‘HbSS’ genotype, the most severe form of SCD. Benefits of zinc supplementation on nutritional indices and anthropometric parameters are well established in literature. Zemel and colleagues (2002) reported on improvement in longitudinal growth (height, 117.4cm ± 10.8 to 124.4cm ± 10.7, p<0.005) and
body weight (20.6 kg ± 4.2 to 23.5 kg ± 5.0, p<0.005) of growing children with SCA when given zinc supplements in a 12-month period.

The role of zinc in decreasing oxidative stress and inflammatory cytokines, as well as increasing anti-inflammatory proteins in sickle cell patients has also been documented (Nwaoguikpe and Braide, 2012). Experimentally, the mineral has also been shown to exhibit high inhibition of sickle cell haemoglobin polymerisation compared to other antioxidants such as vitamin A, E and C. (Nwaoguikpe and Braide, 2012).

2.10.1.3 Folic acid

Folic acid is an essential vitamin needed for the production of red blood cells and the formation of neurotransmitters in the brain. Rich sources of the natural form (folate) include green leafy vegetables such as spinach, legumes, liver etc. As part of routine management of SCD, folic acid is being prescribed in almost all health centres in both developing and developed countries. It has been recommended that diet for sickle cell patients should be high in folate as needed in much as pregnancy (400 to 600 mcg daily) because of the increased production of erythrocytes needed to replace the cells being continuously destroyed and to prevent megaloblastic erythropoiesis (Mahan et al., 2012). There are inconsistencies in literature on the clinical benefits of the supplement on the disease manifestations. Some reports have cited low serum and erythrocyte folate levels in paediatric sickle cell patients and high incidence of megaloblastic anaemia (Van der Dijs et al., 2002). The positive effects of supplementation of the vitamin have also been reported and these include reversal of developmental delay, reduced dactylitis, reduction of homocysteine levels and reduced risk of cardiovascular disease (Al-Yassin et al., 2012). Conversely, others have reported no deficiency in folate, no improvement in haemoglobin status, growth characteristics and infections among patients with SCD after supplementation of the vitamin (Rabb et al., 1983).
2.10.3 Antioxidant vitamins

Antioxidants are compounds that inhibit or scavenge free radicals released during peroxidation reactions. Their deficiency either endogenously or exogenously may lead to oxidative stress (imbalance of oxidants and antioxidants). Oxidative stress is common among patients with SCD due to deficient in antioxidant vitamins and minerals (Allard et al., 1998). Red blood cell membranes of patients with SCD are susceptible to endogenous free-radical damage with clinical manifestations of chronic haemolysis and subsequent anaemia (Nwaoguikpe & Braide, 2012). The production of reactive oxygen species (ROS) can be increased in response to a variety of patho-physiological conditions such as hypoxia, inflammation, infection, dehydration and deficiency in antioxidant vitamins. Amer et al. (2006) reported 20-50% lower levels of reduced glutathione (GSH), the major intracellular scavenger of ROS and 10-30-fold higher production of ROS in sickle cell patients compared to healthy controls (Hb AA) (p<0.005). They further showed that exposure of blood samples of sickle cell patients to antioxidants such as N-acetylcysteine, vitamin C and vitamin E decreased oxidative stress in 2-fold compared to control (p<0.05). This suggests that antioxidant treatment of patients with SCD could reduce oxidative damage to red blood cells especially when administered in combination.

2.10.3.1 Vitamin E

Vitamin E is an important lipid-soluble antioxidant vitamin. The vitamin plays an indispensable role in protecting red blood cell membrane from free radical damage in the body. Rich dietary sources include peanut oil, olive oil, canola oil, mixed nuts, and almonds (Mahan et al., 2012). Naturally, the vitamin is also present in many fruits and foods such as avocados, shrimps, nuts (almonds), broccoli, tomatoes, sunflower seeds, vegetable oil and potatoes. Of all the antioxidant vitamins, it has been the most investigated vitamin in relation to SCD. There have been reports of low circulating level of vitamin E in SCD patients, although there are also contrasting reports of normal levels. Lower levels have been associated with increased
haemolysis, increased susceptibility to infections, and sickling of red blood cells among SCD patients (Essien, 1995).

### 2.10.3.2 Vitamin C

Vitamin C is a nutrient required in very small amounts to perform a range of essential metabolic functions in the body. It is mainly recognised by the role it plays in collagen synthesis which is required for connective tissue formation, as an antioxidant, and the prevention of scurvy. Rich source of the vitamin is mostly from fruits (banana, orange, apple, mango, cherry, pineapple) and vegetables (onion, tomatoes, spinach, broccoli) (Mahan et al., 2012). Deficiency of vitamin C is associated with delayed in wound healing, increased susceptibility to infections, and scurvy characterised by gum bleeding and sore mouth. Daily recommendation for children is 45mg and 60mg for adults. (Chiu et al., 1990). Increased utilisation of the vitamin for disease process may account for its deficiency among patients with SCD. Lower serum levels of vitamin C have been measured in sickle cell patients when compared to normal controls (18.3±9.4µg/108 cells, 30.3±7.5 µg/108 cells; p < 0.01), although dietary intake was adequate. The investigators also concluded that pre-treatment of sickle cells with ascorbic acid protects their membranes against in vitro peroxidative lipid damage (Jain & Williams, 1985). Other supplementation benefits include in vitro inhibition of formation of dense cells and Heinz bodies (denatured Hb) (Ohnishi, Ohnishi, & Ogunmola, 2000). However, it is recommended that supplementation be done with care since vitamin C increases iron absorption which may lead to iron overload and exacerbate disease complications (Mahan et al., 2012).

### 2.10.4 Vitamin A

Vitamin A is one of the fat-soluble vitamins indispensably required for growth and development, normal vision, the expression of selected genes, cell proliferation and differentiation as well as maintaining the integrity of the immune system. Dietary sources include carrot, sweet potato, liver, fish oils, full-cream milk, butter, and organ meat (Shils and Shike, 2006). Deficiency of the vitamin in both healthy and sickle cell children has been associated with higher rates of
morbidity and mortality with poor growth and development (Villamor and Fawzi, 2000). Schall et al., (2004) reported prevalence of suboptimal status of Vitamin A in US sickle cell children and associated it with increased number of hospitalization, poor growth and lower haematological status although mean dietary intake of the vitamin exceeded the RDA. They suggested that Vitamin A supplementation may be beneficial for reducing hospitalizations and improving the quality of lives of people affected with SCD.

2.10.5 Vitamin D

Vitamin D is a fat-soluble vitamin responsible for the maintenance of calcium and phosphate homeostasis and is vital for bone health. It is normally referred to as ‘the sunshine vitamin’ because exposure to sunlight is usually sufficient for its synthesis by the skin. This makes deficiency of the vitamin less a problem in tropical Africa. Deficiency of the vitamin can lead to bone deformities such as rickets in children. It can also result in bone pain, and tenderness (osteomalacia) in adults (Mahan et al., 2012). In reference to sickle cell disease, there is increasing data demonstrating low serum levels of vitamin D among both healthy and sickle cell children mainly in the developed countries where there is less sunshine. Other possibility may be due to decreased dietary intake and in some cases to seasonal variability in food intake (Hyacinth et al., 2010). Rover et al (2008) have reported that about 33% of their sickle cell participants were vitamin D deficient ([25(OH)D <11 mg/mL] and had lower intake. Lal et al (2006) have reported similar findings. SCD patients may benefit from routine vitamin D and calcium supplements, to increase bone mineral density (BMD) and reduce recurrent bone pain. (Hyacinth et al., 2010).

2.10.6 Magnesium

Magnesium is recognised as the second-most abundant (after potassium) intracellular mineral in the body. The adult human body contains about 20 to 28 g of magnesium, of which approximately 26% is found in the muscle, 60% in bone, and the rest in soft tissues and body fluids (Mahan et al., 2012). Aside its role in stabilizing the structure of ATP in ATP-dependent
enzyme catalysed reactions, the macro-mineral is also vital in neuromuscular transmission and activity. Magnesium is found in many foods. Good sources are seeds, nuts, legumes, milled cereal grains, and dark green vegetables. High magnesium intakes are associated with greater bone density (Rude & Gruber, 2004). The mineral is thought to decrease sickle erythrocytes dehydration. Studies have reported inconsistent serum levels of magnesium (Mg) in SCD patients. Low levels of total Mg in red blood cells of sickle cell patients have been associated with increased sickling due to tendency for red blood cell dehydration and hence, increased HbS polymerization (Hyacinth et al., 2010).

The administration of magnesium supplements has been reported to improve haematological indices in patients with SCD, including improvement of red cell hydration shown by reduction of number of dense sickle erythrocytes, absolute reticulocyte count and immature reticulocyte (Goldman et al., 2013). Brousseau and colleagues (2004) have reported that intravenous Mg could significantly decrease the length of hospital stay from approximately five days to an average of three days (p=0.01).

2.11 Nutritional recommendation for patients with SCD

Although there is sufficient evidence that hypermetabolism and increased energy expenditure increase nutrient requirements among sickle cell patients, there are no specific dietary recommendations for these patients. However, it has been suggested that, SCD patients should

- Take in enough water as possible each day, at least 8–10 glasses to prevent dehydration.
- Avoid caffeine-containing drinks, such as energy drinks, cola or coffee and alcohol. These cause frequent urination which results in dehydration.
- Avoid icy products which could cause blood vessels to narrow, causing difficulty in blood flow through vessels, hence pain.
- Consume protein-dense foods with emphasis on plant-sources of proteins such as beans, peas, pulses, lentils, soybeans, groundnuts, cashew, and mushroom
• Increase consumption of food sources of citrulline, a nutrient that promotes blood vessels relaxation and improves oxygen and blood circulation. Some sources include watermelon, milk, legumes, and cucumber.

• Consume more food sources of folate, vitamin B6, B12, and zinc which are involved in red blood cell production and maintenance of their membrane integrity.

• Take Iron-rich foods, such as liver, iron-fortified formula, iron-fortified cereals, and iron-fortified energy with caution due to risk of iron overload as well as vitamin C which increases iron absorption (Mahan et al., 2012).

2.12 Dietary intake
Poor health and nutritional status has been associated with low dietary intake due to lack of essential nutrients. Increased dietary intake may be needed to compensate for the disease process in sickle cell children. In a study by Gray et al (1992) to determine the nutritional status and dietary intake of children with sickle cell disease to confirm deficiencies, although dietary intake of protein and energy was more than controls, the SCD children were leaner and weighed less. Again, although dietary intake of zinc, vitamin A, folic acid, and iron was similar to that of controls and adequate compared to the Recommended Dietary Allowances, lower serum level of zinc and vitamin A were seen among the SCD children.

An important aspect of dietary intake is dietary diversity. According to FAO (2011), dietary diversity is a recommended approach to achieving nutritional requirements and a proxy measure for macro and/or micronutrient adequacy of the diet. Increasing dietary diversity scores (DDS) have been associated with higher BMI of children and lower height for age and weight for height (p≤0.024) in non-sickle cell children (Hooshmand & Udipi, 2013; Olumakaiye, 2013). Children with SCD may benefit from dietary diversity due to reduced dietary intake and increased nutrient requirements.
2.13 Recommended Dietary Allowance (RDA)

Recommended Dietary Allowance (RDA) is the average daily dietary intake level sufficient to meet the nutrient requirement of nearly all (97–98%) healthy individuals in a particular life-stage and gender group (Murphy and Poos, 2002). RDAs have been prepared by the Food and Nutrition Board of the National Research Council of the National Academy of Sciences since 1941. RDAs were first published in 1943 when the U.S. population was recovering from a major economic crisis and World War II where nutrient deficiencies were of much concern (Food and Nutrition Board, 1989). The idea behind was to establish guidelines that would promote optimal health, with the goal of reducing nutrient deficiencies. As food supply and the nutritional needs of the population changed, the intent of the RDAs was adapted to preventing nutrition-related diseases.

RDAs have always being set in relation to gender, age, life phase differences, specific conditions (e.g. growth and pregnancy) and is periodically updated to reflect new knowledge. In disease conditions such as SCD where there is an increased requirement, the RDA or AI (adequate intake) may serve as the basis for adjusting individual recommendations (Yates, 1998). Dieticians and health care professionals should adapt the recommended intake (RDA) to cover the increased demand of nutrients.

Although there are circulating data on hypermetabolism and nutrient compensation for chronic diseases such HIV/AIDS and SCD processes, specific RDAs have not being set for these chronic conditions just as it is done to compensate for the increased requirements for extra nutrients during growth and pregnancy.
Table 2.1 Dietary Reference intakes (DRIs): Recommended Dietary Allowances for selected nutrients, Food and Nutrition Board, Institute of Medicine (IOM, 2004) National Academies

<table>
<thead>
<tr>
<th>Nutrient</th>
<th>Age (years)</th>
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<tr>
<td></td>
<td>1-3</td>
<td>4-8</td>
<td>9-13</td>
<td></td>
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<tr>
<td>Protein (g/kg/d)</td>
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<td>0.95</td>
<td>0.95</td>
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<tr>
<td>Energy (Kcal)</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>males</td>
<td>1046</td>
<td>1742</td>
<td>2279</td>
<td></td>
</tr>
<tr>
<td>females</td>
<td>992</td>
<td>1642</td>
<td>2071</td>
<td></td>
</tr>
<tr>
<td>Carbohydrates (g/d)</td>
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<tr>
<td>Iron (mg/d)</td>
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</tr>
<tr>
<td>Zinc (mg/d)</td>
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<tr>
<td>Magnesium (mg/d)</td>
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<tr>
<td>Calcium (mg/d)</td>
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<td>1300</td>
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<td>Vitamin A (µg/d)</td>
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<tr>
<td>Vitamin B9 (µg/d)</td>
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<td>200</td>
<td>300</td>
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<tr>
<td>Vitamin E (mg/d)</td>
<td>6</td>
<td>7</td>
<td>11</td>
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2.14 Nutritional status

Nutritional status is the balance between the intake of nutrients and the expenditure of these in the processes of growth, reproduction and maintenance of health. Assessment of nutritional status is necessary to monitor growth of children and nutrition-related health consequences, as well as to evaluate interventions. Nutritional status assessment involves anthropometry, biochemical investigations, clinical evaluation, and dietary intake. The monitoring of dietary intake and nutritional status of sickle cell children is an essential requirement for comprehensive care to facilitate early diagnosis of growth failure and implementation of nutritional interventions (Al-Saqladi et al., 2008). Nutrient intake is often compromised in sickle cell
patients due to comorbidities such as painful crises and anorexia and this affects nutritional status. Other reasons such as increased resting energy expenditure, increased energy requirements, and higher basal metabolic rate have also been identified as factors accounting for the poor growth and nutritional status seen in children with SCD (Cox et al., 2011). In a study by Gray and colleagues (1992) to determine the nutritional status and dietary intake of children with sickle cell disease, the sickle cell children were leaner and weighed less despite similar dietary intake of zinc, vitamin A, folic acid and iron and had higher intakes of energy and protein when compared to controls. Cox and colleagues also reported high prevalence of stunting (36.2%) with wasting predicting an increased risk of hospital admissions among urban cohort of Tanzania sickle cell anaemia patients. In Ghana the prevalence of malnutrition among sickle cell children has been found to be 61.3% compared to healthy controls 28.6% (p<0.001) (Osei-Yeboah et al., 2011).

Growth assessments are mainly used to evaluate and monitor the nutritional status of a child with height-for-age, weight-for-age, mid-upper arm circumference and head circumference as indicators of growth for all children. Table 2.0 shows the WHO cut-off points used as reference standards for growth assessment in children.

Table 1.2 Anthropometric Cut-Off for Growth Assessment in Children (Onis, 2006)

<table>
<thead>
<tr>
<th>Z-score</th>
<th>Height-for-Age</th>
<th>Weight-for-Age</th>
<th>BMI-for-Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; - 3SD</td>
<td>Severely Stunted</td>
<td></td>
<td>Severely wasted</td>
</tr>
<tr>
<td>&lt; - 2SD</td>
<td>Stunted</td>
<td>Underweight</td>
<td>Wasted</td>
</tr>
<tr>
<td>&gt; - 1SD to</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>&lt; + 1SD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt; + 1SD</td>
<td>Possible risk of overweight</td>
<td></td>
<td>Possible risk of overweight</td>
</tr>
<tr>
<td>&gt; + 2SD</td>
<td>Overweight</td>
<td></td>
<td>Overweight</td>
</tr>
<tr>
<td>&gt; + 3SD</td>
<td>Obese</td>
<td></td>
<td>Obese</td>
</tr>
</tbody>
</table>
2.15 Anthropometry

Anthropometry is the science of obtaining human body measurements (Wang et al., 2000). It involves obtaining the physical measurements of an individual and relating them to standards. Anthropometry includes a variety of measurements such as weight, height, circumferences, and skinfold thickness. Combination of different measurements or combination of a measurement with other data gives an anthropometric index (e.g. weight for height, height for age, weight for age). These measurements are components of nutrition assessment and are useful for evaluating over-nutrition or under-nutrition. Anthropometry has remained a key component of nutritional status assessment both in children and adults especially when combined with biochemical parameters. Accurate measurements of anthropometry data on children may be a good reflection on their dietary intake, general health status, growth and development over time (McDowell & Statistics, 2005). The increased nutrient requirements and/or poor nutritional status documented in children with SCD may be confirmed using anthropometric indices.
CHAPTER THREE

3.0 METHODOLOGY

3.1 Study Setting

The study was conducted at Princess Marie Louise Children’s Hospital (PML). The hospital popularly known as children’s hospital was established in 1926 and was named after Her Highness Princess Louise Marie, the grand-daughter of Queen Victoria. It is situated in the nation’s capital, Accra, precisely within the Korle-Wokon community. The hospital is a specialized hospital that offers integrated service in the management of childhood illnesses. Major health care services include reproductive and child health, family planning, nutrition rehabilitation service, dental, eye, ear, nose and throat, asthma, HIV clinics, oral rehydration point and sickle cell disease clinics. Departmental units include outpatient department, dispensary, laboratory, medical records, catering, public health and general administration units, as well as a mortuary. The hospital also has a recreational centre for children.

The paediatric sickle cell clinic is one of the speciality clinics at the hospital. The clinic started in 2007 and has about 471 registered sickle cell patients. Children aged 12 years and younger, with confirmed diagnosis of SCD are seen at scheduled routine visits (every 3-4 months), held once a week (Thursdays). It is aimed at keeping the children in steady state and identifying early signs of complications associated with the disease. A total of 25 patients are booked for appointment every week but due to poor attendance about 17 patients are seen in a week.

3.2 Study Design

The study was a hospital-based cross-sectional survey conducted at Princess Marie Louise Children’s Hospital in the Greater Accra Region of Ghana.

3.3 Study Population

The study participants included caregiver’s with children 3-12 years diagnosed of sickle cell disease attending outpatient sickle cell clinic at PML. Recruitment was done as participants awaited medical care.
3.4 Sample Size Determination and Sampling Procedure

Sample size for the study was obtained using the total number of registered sickle cell children at the hospital. With registered number of 471 sickle cell children, the sample size was determined to obtain a percentage of the registered number of SCD children by adopting the following statistical formula for minimum sample size calculation (Yamane, 1967):

\[
n = \frac{N}{1 + N(e)^2}
\]

Where \( n \) = minimum sample size, \( N \) = the sampling frame (i.e. the total number of registered children with sickle cell disease.

\( e \) = the margin of error. Using 8% (0.08) margin of error, a sample size of 118 was obtained.

The sample size was rounded up to 120 to make room for uncompleted questionnaires.

Purposive sampling technique was used to select participants for the study. Participants were purposely selected because they had been diagnosed as having SCD by a medical doctor.

3.4.1 Inclusion and Exclusion criteria

3.4.1.1 Inclusion criteria

Participants were considered eligible if they were

- Aged 3-12 years and diagnosed as having SCD by a medical doctor
- In a steady state and attending sickle cell clinic at outpatient department of PML.

3.4.1.2 Exclusion criteria

Participants were excluded if they were in crisis or had chronic medical conditions such as HIV, tuberculosis, cystic fibrosis other than sickle cell disease.

3.5 Recruitment and training of field Assistants

The study employed two field assistants who were trained for data collection. They were trained on how to administer the research questionnaires first in the English and then in the local language (Twi, Ewe and Ga). They were also trained by a qualified nutritionist on standard
procedures for taking anthropometric measurement. Inconsistencies in the questionnaire administration were identified and corrected during the training session.

3.6 Ethical consideration

Ethical clearances (ECBAS 008/15-16 and GHS-ERC 05/08/15) were obtained from the ethical committee of the College of Basic and Applied Sciences (ECBAS) at the University of Ghana and Ghana Health Service (GHS). Issues regarding confidentiality, anonymity, risks and benefits, freedom to participate and withdraw from the study at any time without any adverse consequences were categorically stated. The researcher explained the purpose of the research to each caregiver with the aid of participant information sheet. The caregivers were then given the volunteer agreement sheet to complete and sign to certify that they had understood and agreed to their child being part of the study. Assents were also obtained from the children. Individual participants were given unique codes to maintain anonymity and to ensure confidentiality.

3.7 Data Collection

Data were collected from 120 participants who had been consented and had assented to be part of the study. The questionnaire captured information on the demographic data of the child, socio-demographic characteristic of the caregiver, clinical information of the child which was either obtained from the caregiver and/or from their medical records. The last part of the questionnaire captured information on the child’s dietary intake (24 hour recall and food frequency questionnaire) and anthropometric parameters as well as the child’s haemoglobin level. The data collection lasted for approximately 3 months (January to March, 2016).

3.7.1 Demographic and clinical data

A semi-structured questionnaire was used to collect demographic information from the caregivers and children. These included child’s date of birth, sex, age, place of residence, educational level etc. Clinical data such as type of genotype, transfusion information, date of
diagnosis and first clinic attendance, number of hospitalization in the past 12 months were obtained from their medical records.

3.7.2 Dietary Assessment

3.7.2.1 Dietary diversity and frequency (24 hours)

A single 24-hour recall was used to collect dietary information on the different number of food groups the child has eaten from within the 24-hour period prior to the interview. Parents and/or children were asked to recall all the foods and drinks eaten the previous day. The weights or quantities of the foods consumed by the children were estimated using household food measures and in instances where caregivers were unable to estimate the quantity of the food consumed with a household measure, it was estimated in cost or amounts and the same foods were purchased from food vendors and then weighed using an Ohaus CS 2000 compact scale. Since the interview was done on weekday (Thursdays), the children were asked to recall the food they ate in school particularly lunch. Where a child was unable to recall the food, the researcher noted the name of the school and estimated the amount served to the children from the school to get an estimate of the child’s intake. This was done mostly for children less than 5 years of age.

3.7.2.2 Food group frequency (for the past seven 7 days)

Information about the number of times a child had eaten from a particular food group during the past seven days was collected using food group frequency questionnaire (FAO, 2011). The nine food groups included: starchy staples, meat and fish, legumes, nuts and seeds, eggs, milk and milk products, vitamin A rich fruits and vegetables, other fruits and vegetables, dark green leafy vegetables, and organ meat. Caregivers were asked to recall the number of days in a week that children ate from each of these nine food groups.

3.7.3 Haemoglobin level determination

Haemoglobin concentrations of the children were determined using haemoglobin meter (URIT-12). The middle finger was used for the sampling. The puncture site was cleaned and wiped dry using alcohol swap (70% alcohol). Using my thumb in a gentle rocking movement, the finger
was pressed lightly from the top knuckle to the tip to stimulate the flow of blood to the sampling point. The lancet was used to prick the side of the finger. The first drop of blood was wiped away with a dry absorbent pad (cotton wool). This stimulated spontaneous blood flow and avoided any tissue fluid that may give false readings. The next blood flow (10 µl) was used for haemoglobin estimation. Dried cotton wool was put at the pricked area to stop further bleeding. Values were recorded in grams per deciliters (g/dl). This test was performed by a trained Biomedical Scientist.

3.7.4 Anthropometric Measurements

The nutritional status of the children was established by measuring their weight and height by the Researcher and trained field assistants. Before taking the measurements, the researcher explained to the caregivers and the children why measurements were taken.

3.7.4.1 Weight measurement

The weight of each child was determined using digital scale (Seca, 874). The child stood alone on the calibrated scale placed on a hard floor. The children were weighed in minimal clothing and bare footed. The scale was re-zeroed before the child stepped on and stood in the centre section of the scale, with feet a little distance apart. The child stood still, but relaxed. The weight was taken and recorded to the nearest 0.1 kg. Measurements were recorded after being taken twice and the average of the two used for data interpretation (WHO, 2008).

3.7.5.2 Height Measurement

A stadiometer with a sliding height rod was used to measure the heights of the children. The stadiometer was placed on the hard floor and the children were asked to remove shoes, hats and any other heavy clothing. The children stood on the baseboard with feet a little distance apart with back of the head, buttocks and legs all touching the upright rod. The child’s head looked
straight forward so that the chin was parallel to the baseboard. To keep the child in this position a hand was placed under the chin. The other hand was used to lower the height rod to be placed on top of the head of the child, compressing the hair. Measurements were read out and recorded in centimetres (cm) to the nearest 0.1 cm. Measurements were recorded after being taken twice and the average of the two used for data interpretation (WHO, 2008).

3.7.5.3 Mid-upper arm circumference measurement (MUAC)

The MUAC of the children were measured by palpating the left shoulder of the child to locate the acromion process and was carefully marked. The arm was then bent at the elbow to make a right angle. One end of a measuring tape was placed on the mark indicated on the acromion process and the tape extended down along the upper arm to the bony structure (tip) of the elbow (olecranon process). The midpoint of the arm was marked with a marker. The arm was then straightened and the tape wrapped around the marked midpoint of the arm without compressing the skin. The reading was taken twice and the average of the used for data interpretation.

3.8 Quality Control Measure

Field assistants were adequately trained on how to administer the questionnaires and how to use anthropometric instruments. The questionnaire was pretested on ten respondents at Ridge hospital, Accra, for consistency and clarity. The weighing scale was calibrated before use. Interviews were conducted in the local languages of caregivers and the questionnaires were cross-checked at the field to ensure that all responses were valid and errors identified were corrected. The data collected was validated by identifying missing values and these were treated as such.

3.9 Data Analysis

Data entry and analyses were done using SPSS version 20.0. Means and standard deviations were used for continuous variables and frequencies and proportions for categorical variables. Ages were categorised into three, <5years, 4-8years and 9-12years to capture vulnerable groups
(<5years). They were not categorised according to Institute of Medicine (IOM) dietary recommendations due to skewness of data (3years=15, 4-8years=90, 9-12years= 15).

The anthropometric measurements for children aged 3-5 years old were analysed using the WHO Anthro v.3.2.2 software, which measures the growth and development of children up to 60 months old. The WHO AnthroPlus v.1.0.4 software was used to analyse data for children older than 60 months (WHO, 2007). Data were presented using the WHO growth indicators of height-for-age (stunting), and BMI-for-age (thinness, severe thinness, overweight, and obesity) at cut-off points indicated in Table 2.0. Children with Z-scores below -2SD of the median reference of height-for-age (HAZ), weight-for-height (WHZ) and weight-for-age (WAZ) were classified as stunted, wasted and underweight respectively. Individual dietary intakes were converted to nutrients using the Ghanaian food composition table and compared with the Recommended Dietary Allowance (RDA) for healthy children (IOM, 2006).

Anaemia was classified for children below and above 5 years as Hb <11.0 g/dl and 11.5 g/dl respectively (WHO, 2011). Severity of illness was classified as severe, moderate and mild cases. Disease severity scores were mainly based on the past one year. Participants were classified as ‘severe’ if they were severely anaemic (Hb<7.0 g/dl), had two or more painful episodes and SCD related admissions in the past one year. They were classified as ‘moderate’ if they did not meet severe criteria but had Hb less than 8g/dl, one painful episode and SCD related admission in the past one year (Logan et al., 2002, Zemel, 2007, Mandese et al., 2016). The remaining participants were classified as mild cases.

Chi-square test was used to determine the association between disease severity, dietary intake and nutritional status. After adjusting for energy, nutrients were compared between age groups using ANCOVA. When differences were detected, post hoc analysis was done to detect were the difference existed between the groups. Multiple logistic regression was used to determine demographic and clinical factors associated with nutritional deficits; stunting and underweight.
CHAPTER FOUR

4.0 RESULTS

4.1 Socio-demographic characteristics

The study involved 120 children with 53.3% being males (Table 4.1). The mean age of the children was 5.9±2.2 years. The ages were categorised into three groups; <5years (36.7%), 5-8 years (50.8%), and 9-12 years (12.5%). More than half (60.0%) of the children were preschoolers while the rest (40.0%) had primary education. More than a third (38.3%) of the children were Akans and a similar proportion (32.5%) belonged to the Ga tribe. Almost all the participants were Christians (93.3%) with few (6.7%) in the Islamic religion. More than half (65.0%) of the children were staying with both parents with few (4.2%) staying with other caregivers who were either grandmothers or grandfathers.

Caregivers of the participants were mostly traders (55.8%) and artisans (46.3%) with few of them (3.3%) not employed. In relation to educational background of caregivers, few of the caregivers were educated to the tertiary level (5.0%) with the rest either having no formal education (10.8%), with primary education (31.7%) or secondary/vocational (19.2%). Although more than half (66.7%) of the caregivers were married, more than a tenth (15.8%) had divorced. Details on participants’ characteristics are presented in Table 4.1 below.
Table 4.1 Socio-demographic characteristics of participants

<table>
<thead>
<tr>
<th>Variable</th>
<th>Number of respondents (N)</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child characteristics</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>64</td>
<td>53.3</td>
</tr>
<tr>
<td>Female</td>
<td>56</td>
<td>46.7</td>
</tr>
<tr>
<td>Age (5.9±2.2 years)*</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;5</td>
<td>44</td>
<td>36.7</td>
</tr>
<tr>
<td>5-8</td>
<td>61</td>
<td>50.8</td>
</tr>
<tr>
<td>9-12</td>
<td>15</td>
<td>12.5</td>
</tr>
<tr>
<td>Educational level</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pre-School</td>
<td>72</td>
<td>60.0</td>
</tr>
<tr>
<td>Primary</td>
<td>48</td>
<td>40.0</td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ga</td>
<td>39</td>
<td>32.5</td>
</tr>
<tr>
<td>Ewe</td>
<td>24</td>
<td>20.0</td>
</tr>
<tr>
<td>Akan</td>
<td>46</td>
<td>38.3</td>
</tr>
<tr>
<td>Northerner</td>
<td>11</td>
<td>9.2</td>
</tr>
<tr>
<td>Religion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Christian</td>
<td>112</td>
<td>93.3</td>
</tr>
<tr>
<td>Islam</td>
<td>8</td>
<td>6.7</td>
</tr>
<tr>
<td>Nationality</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ghanaian</td>
<td>118</td>
<td>98.3</td>
</tr>
<tr>
<td>Other^a</td>
<td>2</td>
<td>1.7</td>
</tr>
<tr>
<td>Guardian</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Both parents</td>
<td>78</td>
<td>65.0</td>
</tr>
<tr>
<td>Single Parent</td>
<td>37</td>
<td>30.8</td>
</tr>
<tr>
<td>Other^b</td>
<td>5</td>
<td>4.2</td>
</tr>
<tr>
<td><strong>Caregivers characteristics</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Educational level</td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>13</td>
<td>10.8</td>
</tr>
<tr>
<td>Primary</td>
<td>38</td>
<td>31.7</td>
</tr>
<tr>
<td>JHS</td>
<td>40</td>
<td>33.3</td>
</tr>
<tr>
<td>SHS/Vocational</td>
<td>23</td>
<td>19.2</td>
</tr>
<tr>
<td>Tertiary</td>
<td>6</td>
<td>5.0</td>
</tr>
<tr>
<td>Occupation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trading</td>
<td>67</td>
<td>55.8</td>
</tr>
<tr>
<td>Artisan</td>
<td>46</td>
<td>38.3</td>
</tr>
<tr>
<td>Government employee</td>
<td>3</td>
<td>2.5</td>
</tr>
<tr>
<td>Unemployed</td>
<td>4</td>
<td>3.3</td>
</tr>
<tr>
<td>Marital Status</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Single</td>
<td>21</td>
<td>17.5</td>
</tr>
<tr>
<td>Married</td>
<td>80</td>
<td>66.7</td>
</tr>
<tr>
<td>Widowed/Divorced</td>
<td>19</td>
<td>15.8</td>
</tr>
</tbody>
</table>

*mean±SD, other^a refers to Nigerian and Togolese, other^b refers to grandparents; JHS-Junior high school; SHS-Senior High School;
4.2 Clinical Information of participants

Most of the clinical information was obtained from patients’ medical records. Majority of the patients (71.7%) were of the SS genotype, the most severe form of SCD (Table 4.2). Other genotypes recorded were SC (27.5%) and SF (0.8%). Most of the caregivers of patients (74.2%) received some form of nutrition education at the health centre. Some of the participants (4.3%) reported visiting the clinic for the first time. Patients were diagnosed of SCD before age one (10%) and as late as from five years (5.8%). Almost half (42.5%) of the patients had been on admission and the main reasons for admissions were vaso-occlusive episodes (92.2%) and anaemia (7.8%). Severity of illness was classified as mild, moderate and severe. Majority of the children (76.7%) had mild illness.

Table 4.2: Clinical information of participants

<table>
<thead>
<tr>
<th>Variable</th>
<th>Number (N)</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sickle cell type</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SS</td>
<td>86</td>
<td>71.7</td>
</tr>
<tr>
<td>SC</td>
<td>33</td>
<td>27.5</td>
</tr>
<tr>
<td>SF</td>
<td>1</td>
<td>0.8</td>
</tr>
<tr>
<td>Nutrition education</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>89</td>
<td>74.2</td>
</tr>
<tr>
<td>No</td>
<td>27</td>
<td>22.5</td>
</tr>
<tr>
<td>Othera</td>
<td>4</td>
<td>3.3</td>
</tr>
<tr>
<td>Age of diagnosis (yrs)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;1</td>
<td>12</td>
<td>10</td>
</tr>
<tr>
<td>1-2</td>
<td>61</td>
<td>50.8</td>
</tr>
<tr>
<td>3-4</td>
<td>40</td>
<td>33.3</td>
</tr>
<tr>
<td>≥5</td>
<td>7</td>
<td>5.8</td>
</tr>
<tr>
<td>Admission past one year</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>51</td>
<td>42.5</td>
</tr>
<tr>
<td>No</td>
<td>69</td>
<td>57.5</td>
</tr>
<tr>
<td>Reason for Admission</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vaso-occlusive crisis</td>
<td>47</td>
<td>92.2</td>
</tr>
<tr>
<td>Anaemia</td>
<td>4</td>
<td>7.8</td>
</tr>
<tr>
<td>Severity of illness</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>92</td>
<td>76.7</td>
</tr>
<tr>
<td>Moderate</td>
<td>8</td>
<td>6.7</td>
</tr>
<tr>
<td>Severe</td>
<td>20</td>
<td>16.7</td>
</tr>
</tbody>
</table>

Othera refers to participants who were on first visit. Severity of illness (Severe: Hb<7.0g/dl plus ≥ 2 painful episodes and SCD related admissions in the past one year; moderate Hb<8.0g/dl plus one painful episode and SCD related admissions in the past one year; the remaining children were classified as ‘mild’).
4.3 Dietary intake of energy and nutrient levels

Presented in Table 4.3 are the mean energy and nutrient intake levels of participants from a single 24 hour dietary recall. The mean energy and protein intake among the children were 1347.9±606.0 Kcal and 3.0±1.9 g/kg respectively. Micronutrient intake levels were: iron (11.3±6.1 mg), folate (116.4±69.5 µg), vitamin C (82.9±194.6 mg), vitamin A (543.2±587.3 µg/RE), vitamin E (1.1±3.5mg), zinc (4.3±2.5 mg), magnesium (63.5±30.4 mg), and calcium (167.7±98.9 mg).

Nutrients were compared between the age groups after adjusting for energy intakes. There were significant differences in the mean intake of protein (p=0.00), folate (p=0.04) and magnesium (p=0.04) among the age groups. Intake of protein decreased significantly with increase in age while magnesium intake increased significantly with increase in age. All other nutrients did not differ significantly between the age groups (p>0.05). Further analysis (post hoc; LSD) showed significant differences in protein intake between <5 years (3.6±0.2 g/kg) and 5-8 years (2.9±0.2 g/kg) (p=0.04) as well as <5 years and 9-12 years (2.1±0.4 g/kg) (p=0.00). For folate, significant difference was found between children <5 years (93.3±10.3 µg) and 5-8 years (126.6±8.7 µg), p=0.02, as well as between <5 years and 9-12 years (140.7±17.9 µg), p=0.02. Further analysis also showed significant difference in mean intake of magnesium between children <5 years (55.6±4.4 mg) and 9-12 years (74.9±7.6 mg) (p=0.03).
Table 4.3: Dietary intake of energy and selected nutrients by age group

<table>
<thead>
<tr>
<th>Nutrients/day</th>
<th>Total (N=120)</th>
<th>&lt;5</th>
<th>5-8</th>
<th>9-12</th>
</tr>
</thead>
<tbody>
<tr>
<td>Energy (Kcal)</td>
<td>1347.9±606.0</td>
<td>1288.1±699.3</td>
<td>1305.1±448.9</td>
<td>1697.8±778.3</td>
</tr>
<tr>
<td>Protein (g/kg)</td>
<td>3.0±1.9</td>
<td>3.6±0.2&lt;sup&gt;c&lt;/sup&gt;</td>
<td>2.9±0.2&lt;sup&gt;d&lt;/sup&gt;</td>
<td>2.1±0.4&lt;sup&gt;d&lt;/sup&gt;</td>
</tr>
<tr>
<td>Iron (mg)</td>
<td>11.3±6.1</td>
<td>12.0±0.5</td>
<td>11.0±0.4</td>
<td>10.6±0.8</td>
</tr>
<tr>
<td>Folate (µg)</td>
<td>116.4±69.5</td>
<td>93.3±10.3&lt;sup&gt;e&lt;/sup&gt;</td>
<td>126.6±8.7&lt;sup&gt;f&lt;/sup&gt;</td>
<td>140.7±17.9&lt;sup&gt;f&lt;/sup&gt;</td>
</tr>
<tr>
<td>Vitamin B12 (µg)</td>
<td>1.1±1.1</td>
<td>0.9±0.2</td>
<td>1.2±0.1</td>
<td>1.4±0.3</td>
</tr>
<tr>
<td>Vitamin C (mg)</td>
<td>82.9±194.6</td>
<td>76.7±26.5</td>
<td>70.2±22.5</td>
<td>86.6±46.2</td>
</tr>
<tr>
<td>Vitamin A (µg/RE)</td>
<td>543.2±587.3</td>
<td>503.2±87.2</td>
<td>615.9±73.9</td>
<td>365.3±152.3</td>
</tr>
<tr>
<td>Vitamin E (mg)</td>
<td>1.1±3.5</td>
<td>1.6±0.5</td>
<td>1.0±0.4</td>
<td>0.2±0.9</td>
</tr>
<tr>
<td>Zinc (mg/d)</td>
<td>4.3±2.5</td>
<td>4.2±0.3</td>
<td>4.3±0.3</td>
<td>4.6±0.6</td>
</tr>
<tr>
<td>Magnesium (mg)</td>
<td>63.5±30.4</td>
<td>55.5±4.4&lt;sup&gt;e&lt;/sup&gt;</td>
<td>66.5±3.7</td>
<td>74.9±7.6&lt;sup&gt;h&lt;/sup&gt;</td>
</tr>
<tr>
<td>Calcium (mg)</td>
<td>167.7±98.9</td>
<td>143.3±14.0</td>
<td>181.8±11.9</td>
<td>181.8±24.5</td>
</tr>
</tbody>
</table>

Total Nutrients, Energy (Means±SD); across the age groups, values are presented as adjusted means and standard error. Covariates appearing in the model include energy (1347.9) for nutrients comparison. Means on the same row with different letters are significantly different. RE= Retinol equivalent, *p*-value<0.05, significance in the Ancova model.

### 4.3.1 Dietary intakes compared with recommendations

Nutrient levels were compared with Institute of Medicine (IOM) recommendations for recommended daily allowance (RDA). Recommendations are age specific (1-3 years, 4-8 years, 9-13 years etc.) hence the children were regrouped (age 3, 4-8 years and 9-12 years). Children aged three consumed more than 100% of the RDA for energy, protein, iron, vitamin A, and zinc while less than 50% of the RDA for vitamin E and calcium were consumed (Table 4.4). At this age all the children met RDA for protein whiles none met RDA for vitamin E and calcium. Almost all the children (96.7%) within the age of 4-8 years met RDA for protein but not for vitamin E and calcium. Most of the children (86.7%) within the age of 9-12 years met RDA for protein but none met RDA for folate, vitamin E, magnesium and calcium.
Table 4.4: Dietary data compared to age specific recommended dietary allowance (RDA)

<table>
<thead>
<tr>
<th>Age group (years)</th>
<th>Nutrients/day</th>
<th>Mean ±SD</th>
<th>RDA*</th>
<th>% RDA</th>
<th>N (%) meeting RDA</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 (n=15)</td>
<td>Energy (Kcal)</td>
<td>1439.2±800.2</td>
<td>1046</td>
<td>162.3±74.3</td>
<td>12 (80.0)</td>
</tr>
<tr>
<td></td>
<td>Protein (g/kg)</td>
<td>3.9±2.4</td>
<td>1.05</td>
<td>273.6±187.9</td>
<td>15 (100)</td>
</tr>
<tr>
<td></td>
<td>Iron (mg)</td>
<td>13.7±11.4</td>
<td>7</td>
<td>195.4±88.2</td>
<td>13 (86.7)</td>
</tr>
<tr>
<td></td>
<td>Folate (µg)</td>
<td>98.5±52.7</td>
<td>150</td>
<td>94.6±43.4</td>
<td>7 (46.7)</td>
</tr>
<tr>
<td></td>
<td>Vitamin B12 (µg)</td>
<td>1.0±0.7</td>
<td>0.9</td>
<td>125.6±118.7</td>
<td>6(40.0)</td>
</tr>
<tr>
<td></td>
<td>Vitamin C (mg)</td>
<td>15.7±11.7</td>
<td>15</td>
<td>97.9±31.1</td>
<td>9 (60.0)</td>
</tr>
<tr>
<td></td>
<td>Vitamin A (µg/RE)</td>
<td>568.2±508.2</td>
<td>300</td>
<td>313.9±395.0</td>
<td>4 (26.7)</td>
</tr>
<tr>
<td></td>
<td>Vitamin E (mg)</td>
<td>3.1±9.8</td>
<td>6</td>
<td>14.3±7.8</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td></td>
<td>Zinc (mg/d)</td>
<td>3.9±3.3</td>
<td>3</td>
<td>177.3±86.2</td>
<td>12 (80.0)</td>
</tr>
<tr>
<td></td>
<td>Magnesium (mg)</td>
<td>47.8±22.1</td>
<td>80</td>
<td>98.8±45.2</td>
<td>6 (40)</td>
</tr>
<tr>
<td></td>
<td>Calcium (mg)</td>
<td>141.9±69.8</td>
<td>700</td>
<td>39.9±21.3</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>4-8 (n=90)</td>
<td>Energy (Kcal)</td>
<td>1274.4±517.3</td>
<td>1742</td>
<td>73.2±29.5</td>
<td>13 (14.4)</td>
</tr>
<tr>
<td></td>
<td>Protein (g/kg)</td>
<td>2.9±1.9</td>
<td>0.95</td>
<td>120.5±108.1</td>
<td>87 (96.7)</td>
</tr>
<tr>
<td></td>
<td>Iron (mg)</td>
<td>10.6±4.6</td>
<td>10</td>
<td>105.6±46.0</td>
<td>47 (52.2)</td>
</tr>
<tr>
<td></td>
<td>Folate (µg)</td>
<td>114.8±72.1</td>
<td>200</td>
<td>57.4±35.8</td>
<td>9 (10)</td>
</tr>
<tr>
<td></td>
<td>Vitamin B12 (µg)</td>
<td>1.1±1.1</td>
<td>1.2</td>
<td>94.4±89.1</td>
<td>58 (48.3)</td>
</tr>
<tr>
<td></td>
<td>Vitamin C (mg)</td>
<td>28.8±22.5</td>
<td>25</td>
<td>115.3±89.4</td>
<td>45 (50.0)</td>
</tr>
<tr>
<td></td>
<td>Vitamin A (µg/RE)</td>
<td>556.4±613.8</td>
<td>400</td>
<td>139.1±152.6</td>
<td>41 (45.6)</td>
</tr>
<tr>
<td></td>
<td>Vitamin E (mg)</td>
<td>0.9±0.6</td>
<td>7</td>
<td>12.2±9.0</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td></td>
<td>Zinc (mg/d)</td>
<td>4.2±2.3</td>
<td>5</td>
<td>83.8±44.9</td>
<td>62 (68.9)</td>
</tr>
<tr>
<td></td>
<td>Magnesium (mg)</td>
<td>63.7±29.4</td>
<td>130</td>
<td>49.0±22.5</td>
<td>2 (2.2)</td>
</tr>
<tr>
<td></td>
<td>Calcium (mg)</td>
<td>166.6±101.3</td>
<td>1000</td>
<td>20.8±12.6</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td>9-12 (n=15)</td>
<td>Energy (Kcal)</td>
<td>1697.8±778.3</td>
<td>2279</td>
<td>74.5±32.9</td>
<td>4(26.7)</td>
</tr>
<tr>
<td></td>
<td>Protein (g/kg)</td>
<td>2.9±1.9</td>
<td>0.95</td>
<td>102.4±100.6</td>
<td>13 (86.7)</td>
</tr>
<tr>
<td></td>
<td>Iron (mg)</td>
<td>13.7±6.2</td>
<td>8</td>
<td>170.9±74.6</td>
<td>11 (73.3)</td>
</tr>
<tr>
<td></td>
<td>Folate (µg)</td>
<td>141.8±65.1</td>
<td>300</td>
<td>47.3±20.9</td>
<td>0 (0.0)</td>
</tr>
<tr>
<td></td>
<td>Vitamin B12 (µg)</td>
<td>1.4±1.1</td>
<td>1.8</td>
<td>62.8±59.4</td>
<td>8 (53.3)</td>
</tr>
<tr>
<td></td>
<td>Vitamin C (mg)</td>
<td>54.7±41.7</td>
<td>45</td>
<td>165.9±101.2</td>
<td>2 (13.3)</td>
</tr>
<tr>
<td></td>
<td>Vitamin A (µg/RE)</td>
<td>439.9±514.9</td>
<td>600</td>
<td>73.3±82.9</td>
<td>4 (26.7)</td>
</tr>
</tbody>
</table>
RDA=Recommended dietary allowance; RDAs are set to meet the needs of almost all (97 to 98 percent) individuals in a specific age and gender group. (www.nap.edu)

Results showed that meeting nutrient requirements declined with increase in age (Figure 4.1). The children aged three met RDA for energy and more than half (60.0%) of the ten nutrients that were investigated. Children between 4-8 years and 9-12 years met RDA for only four (protein, iron, Vitamin C and Vitamin A) and three (protein, iron and vitamin C) of the nutrients respectively.

4.3.2 Food consumption from food groups

Consumption of foods from the food groups based on the 24hr dietary recall showed that all the children (100%) consumed food from starchy food group followed by meat and fish (79.2%), legumes and nuts (44.2%), eggs (40.8%), milk and milk products (35%), vitamin A rich fruits and vegetables (29.8%). None of the children consumed organ meat (Figure, 4.2).
Figure 4.2 Percentage consumption of food from the food groups

4.3.3 Dietary Diversity Score (DDS)

Dietary diversity score was calculated from the single 24-hour dietary recall using nine food groups (FAO, 2011). The mean dietary diversity score was 3.7±1.27. The children consumed between one and seven food groups with more than a third (40%) consuming food from three food groups out of the nine food groups. Dietary diversity score was classified as ‘low’, ‘medium’ and ‘high’ when a child consumed less than three, four or five and six or more food groups respectively (FAO, 2011). More than half (53%) of the children had low dietary diversity score while 10% of them had high dietary diversity score (Figure 4.3).
Food consumed in the past 7-days prior to the interview was captured using food frequency questionnaire. Table 4.5 below shows the number of days children ate from the seven food groups. Starchy staples was the most consumed food group and was consumed in almost all the days of the week with an average consumption of $6.8 \pm 0.3$ days, while organ meat was the least consumed food group ($0.1 \pm 0.3$ days).

### Table 4.5: Frequency of consumption from food groups in the past 7- days

<table>
<thead>
<tr>
<th>Food groups</th>
<th>All (N=120)</th>
<th>&lt;5</th>
<th>5-8</th>
<th>9-12</th>
</tr>
</thead>
<tbody>
<tr>
<td>Starchy staples</td>
<td>6.8±0.3</td>
<td>6.9±0.4</td>
<td>6.9±0.3</td>
<td>6.9±0.3</td>
</tr>
<tr>
<td>Meat and fish</td>
<td>6.2±0.9</td>
<td>6.2±1.0</td>
<td>6.2±0.9</td>
<td>6.3±1.1</td>
</tr>
<tr>
<td>Legumes, nuts and seeds</td>
<td>1.8±1.1</td>
<td>1.8±1.2</td>
<td>1.8±0.9</td>
<td>2.1±1.3</td>
</tr>
<tr>
<td>Eggs</td>
<td>1.7±1.1</td>
<td>1.7±1.1</td>
<td>1.7±1.1</td>
<td>2.4±1.2</td>
</tr>
<tr>
<td>Milk and milk products</td>
<td>1.9±1.5</td>
<td>1.9±1.5</td>
<td>1.8±1.4</td>
<td>1.9±2.0</td>
</tr>
<tr>
<td>Vitamin A rich foods and vegetables</td>
<td>2.5±1.2</td>
<td>2.7±1.2</td>
<td>2.5±1.2</td>
<td>2.2±1.4</td>
</tr>
<tr>
<td>Other fruits and vegetables</td>
<td>2.8±1.4</td>
<td>2.9±1.3</td>
<td>2.8±1.4</td>
<td>2.6±1.8</td>
</tr>
<tr>
<td>Dark green leafy vegetables</td>
<td>2.7±1.1</td>
<td>2.7±1.0</td>
<td>2.7±1.2</td>
<td>2.3±1.4</td>
</tr>
<tr>
<td>Organ meat</td>
<td>0.1±0.3</td>
<td>0.1±0.3</td>
<td>0.1±0.2</td>
<td>0.1±0.3</td>
</tr>
</tbody>
</table>

*Mean±SD, tAnova for continuous variables, statistical significance is at p<0.05.
4.4 Disease severity

Severity of illness was classified as severe, moderate and mild (Table 4.2). Participants were classified as ‘severe’ if they were severely anaemic (Hb<7.0g/dl), had two or more painful episodes and SCD related admissions in the past one year. They were classified as ‘moderate’ if they did not meet severe criteria but had Hb less than 8g/dl, one painful episode and SCD related admission in the past one year. The remaining participants were classified as mild cases. Majority of the children (76.7%) had mild cases with few (6.7%) moderate cases and 16.7% severe cases.

4.4.1 Energy and Nutrient intake by disease severity

Presented in Table 4.6 is the energy intakes and nutrients by severity of disease. Nutrient intakes did not differ significantly by severity of disease among the study participants except for vitamin B12 (p=0.02). Post hoc test (Scheffe) showed significant difference between mild (1.0±0.9 µg) and moderate (2.1±1.2 µg) form of disease severity (p=0.02).

Table 4.6: Mean intakes of energy and nutrients by disease severity

<table>
<thead>
<tr>
<th>Nutrients/day</th>
<th>Mild (n=92)</th>
<th>Moderate (n=8)</th>
<th>Severe (n=20)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Energy (Kcal)</td>
<td>1384.6±636.9</td>
<td>1243.4±622.6</td>
<td>1221.3±432.2</td>
<td>0.48</td>
</tr>
<tr>
<td>Protein (g/kg)</td>
<td>3.0±2.0</td>
<td>3.0±1.9</td>
<td>2.9±1.9</td>
<td>0.98</td>
</tr>
<tr>
<td>Iron (mg)</td>
<td>11.9±6.5</td>
<td>9.8±4.0</td>
<td>9.1±5.7</td>
<td>0.23</td>
</tr>
<tr>
<td>Folate (µg)</td>
<td>115.4±64.3</td>
<td>130.9±118.2</td>
<td>113.7±72.3</td>
<td>0.82</td>
</tr>
<tr>
<td>Vitamin B12(µg)</td>
<td>1.0±0.9&lt;sup&gt;a&lt;/sup&gt;</td>
<td>2.1±1.2&lt;sup&gt;b&lt;/sup&gt;</td>
<td>1.2±1.3&lt;sup&gt;b&lt;/sup&gt;</td>
<td>&lt;sup&gt;0.02&lt;/sup&gt;</td>
</tr>
<tr>
<td>Vitamin C (mg)</td>
<td>86.4±196.6</td>
<td>41.3±42.2</td>
<td>33.8±32.3</td>
<td>0.41</td>
</tr>
<tr>
<td>Vitamin A (RE)</td>
<td>541.2±579.3</td>
<td>726.7±687.6</td>
<td>479.0±599.7</td>
<td>0.60</td>
</tr>
<tr>
<td>Vitamin E (mg)</td>
<td>1.2±3.9</td>
<td>0.7±0.4</td>
<td>0.8±5.9</td>
<td>0.88</td>
</tr>
<tr>
<td>Zinc (mg)</td>
<td>4.5±2.6</td>
<td>3.7±1.7</td>
<td>3.6±2.3</td>
<td>0.31</td>
</tr>
<tr>
<td>Magnesium (mg)</td>
<td>63.3±30.5</td>
<td>59.8±31.0</td>
<td>66.2±30.9</td>
<td>0.87</td>
</tr>
<tr>
<td>Calcium (mg)</td>
<td>167.4±100.4</td>
<td>154.1±61.9</td>
<td>174.1±107.3</td>
<td>0.89</td>
</tr>
</tbody>
</table>

<sup>1</sup>Anova for continuous variable; Post hoc (Scheffe’s test), Means on the same row with different letters are significantly different from each other.
4.4.2 Relationship between severity of disease and dietary diversity

Dietary diversity was used as index of dietary intake. Chi-square test showed no significant association (p=0.33) between dietary diversity classification and severity of disease (Figure 4.4).

*Figure 4.4 Association between dietary diversity and disease severity*
4.5: Anthropometric data of participants

Anthropometric data from the survey were compared among the age groups. The mean height-for-age Z-score (HAZ) for all the children was -0.9 ± 1.4 while the mean weight-for-age (WAZ) was -0.8 ± 1.1 (Table 4.7). Of the 120 children, WAZ was computed for 115 children since the WHO anthroplus (V1.04) does not compute WAZ for a child whose age is above 120 completed months. Weight for height Z-score (WHZ) was computed for children less than 5 years while BMI-for-age Z-score was computed for children 5 years and above. The mean BMI-for-age was -0.7±1.1. There was a significant difference for BMI-for-age (p=0.04) between children aged 5-8 years (-0.6±0.9) and 9-12 years (-1.2±1.2).

The prevalence of stunting, underweight, wasting and thinness among the children were 25.8%, 20.0%, 6.8% and 15.8% respectively (Figure 4.4). Significant difference in thinness was observed between the two age groups (5-8 years and 9-12 years) compared (p=0.01).

Anthropometric indices did not differ significantly between males and females (p>0.05).

The study children were categorized into malnourished and well-nourished based on whether a child was stunted, wasted, thin, overweight or underweight. A child was classified as malnourished if he or she had any of these nutritional deficits, and a child was classified well-nourished if these were absent. Figure 4.4 below shows that 38% of the children were malnourished.

Table 4.8 describes the consumption of foods from different food groups based on the nutritional status of the participants. Well-nourished children significantly consumed more meat and fish (p=0.03) as well as milk and milk products (p= 0.02) than malnourished children.
Table 4.7: Nutritional status of children by sex and age group

<table>
<thead>
<tr>
<th>Indicators</th>
<th>Total</th>
<th>&lt;5 (n=44)</th>
<th>5-8 (n=61)</th>
<th>9-12 (n=15)</th>
<th>p-value¹</th>
<th>Males</th>
<th>Females</th>
<th>p-value²</th>
</tr>
</thead>
<tbody>
<tr>
<td>WAZ</td>
<td>0.8±1.1*</td>
<td>-0.8±1.1</td>
<td>0.9±1.1</td>
<td>-0.9±1.5</td>
<td>0.91</td>
<td>-0.9±1.2</td>
<td>-0.8±0.9</td>
<td>0.93</td>
</tr>
<tr>
<td>HAZ</td>
<td>0.9±1.4</td>
<td>0.9±1.4</td>
<td>0.8±1.3</td>
<td>-0.8±1.4</td>
<td>0.73</td>
<td>-0.7±1.4</td>
<td>-0.9±1.3</td>
<td>0.32</td>
</tr>
<tr>
<td>WHZ</td>
<td>0.4±1.0</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-0.6±1.3</td>
<td>-0.2±0.8</td>
<td>0.26</td>
</tr>
<tr>
<td>BMI-for-age</td>
<td>0.7±1.1</td>
<td>-</td>
<td>0.6±0.9</td>
<td>-1.2±1.2</td>
<td>0.04</td>
<td>-0.6±0.9</td>
<td>-0.9±1.2</td>
<td>0.39</td>
</tr>
</tbody>
</table>

* Means±SD, ¹ Anova for continuous variables (WAZ, HAZ); ² Independent t-test for BMI-for-age and sex.

Figure 4.5 Anthropometric indices of the children
Figure 4.6 Pie-chart showing percentage of malnourished and well-nourished children

Table 4.8: Consumption of food from food groups based on nutritional status

<table>
<thead>
<tr>
<th>Food groups</th>
<th>Well nourished</th>
<th>Malnourished</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Starchy staples</td>
<td>6.9±0.4</td>
<td>6.9±0.3</td>
<td>0.59</td>
</tr>
<tr>
<td>Meat and fish</td>
<td>6.4±0.9</td>
<td>5.9±1.1</td>
<td>0.03</td>
</tr>
<tr>
<td>Legumes, nuts and seeds</td>
<td>1.9±1.1</td>
<td>1.8±0.9</td>
<td>0.61</td>
</tr>
<tr>
<td>Eggs</td>
<td>1.8±1.3</td>
<td>1.7±0.8</td>
<td>0.59</td>
</tr>
<tr>
<td>Milk and milk products</td>
<td>2.1±1.5</td>
<td>1.5±1.4</td>
<td>0.02</td>
</tr>
<tr>
<td>Vitamin A rich foods and vegetables</td>
<td>2.4±1.2</td>
<td>2.6±1.3</td>
<td>0.37</td>
</tr>
<tr>
<td>Other fruits and vegetables</td>
<td>2.7±1.4</td>
<td>3.0±1.4</td>
<td>0.14</td>
</tr>
<tr>
<td>Dark green leafy vegetables</td>
<td>2.7±1.1</td>
<td>2.7±1.1</td>
<td>0.81</td>
</tr>
<tr>
<td>Organ meat</td>
<td>0.1±0.3</td>
<td>0.1±0.15</td>
<td>0.06</td>
</tr>
</tbody>
</table>

*Independent t-test for continuous variables, statistical significance is at p<0.05.
4.6 Relationship between severity of disease and nutritional status

Disease severity classification (mild, moderate and severe) was cross-tabulated with nutritional status (malnourished and well-nourished) and this showed no significant association (p=0.24).

This is presented in Figure 4.6 below

![Figure 4.6 Relationship between severity of disease and nutritional status](image)

**Figure 4.7 Relationship between severity of SCD and nutritional status**

However, when severity of illness was cross-tabulated with the individual anthropometric indices, a significant association was observed between underweight and disease severity (p=0.04) as well as wasting (p=0.02) (Table 4.9).

<table>
<thead>
<tr>
<th>Anthropometric indices</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
<th>Total</th>
<th>p-value¹</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Stunting</strong> Normal</td>
<td>71(59.2)</td>
<td>6 (5.0)</td>
<td>12 (10.0)</td>
<td>89 (74.2)</td>
<td>0.28</td>
</tr>
<tr>
<td>Stunted</td>
<td>2 (17.5)</td>
<td>2 (1.7)</td>
<td>8 (6.7)</td>
<td>31 (25.8)</td>
<td></td>
</tr>
<tr>
<td><strong>Underweight</strong> Normal</td>
<td>74 (64.3)</td>
<td>6 (5.2)</td>
<td>12 (10.4)</td>
<td>92 (80.0)</td>
<td><strong>0.04</strong></td>
</tr>
<tr>
<td>Underweight</td>
<td>13 (11.3)</td>
<td>2 (1.7)</td>
<td>8 (7.0)</td>
<td>23 (20.0)</td>
<td></td>
</tr>
<tr>
<td><strong>Wasting</strong> Normal</td>
<td>34 (77.3)</td>
<td>3 (6.8)</td>
<td>4 (9.1)</td>
<td>41 (93.2)</td>
<td><strong>0.02</strong></td>
</tr>
<tr>
<td>Wasted</td>
<td>1 (2.3)</td>
<td>0 (0.0)</td>
<td>2 (4.5)</td>
<td>3 (6.8)</td>
<td></td>
</tr>
<tr>
<td><strong>BMI-for-age</strong> Normal</td>
<td>48 (63.2)</td>
<td>4 (5.3)</td>
<td>12 (15.8)</td>
<td>64 (84.2)</td>
<td>0.96</td>
</tr>
<tr>
<td>Thin</td>
<td>9 (11.8)</td>
<td>1 (1.3)</td>
<td>2 (2.6)</td>
<td>12 (15.8)</td>
<td></td>
</tr>
</tbody>
</table>

¹Pearson chi-square test for categorical variables. Statistical significant is at p-value < 0.05.
4.7 Prevalence of anaemia among the children

Haemoglobin level of the children varied from 5.3 to 11.8 g/dl with mean a of 7.8 ±1.39 g/dl. From the results, almost all the children (98.3%) were anaemic (Table 4.8). Severe form of anaemia was mostly found among children aged 5-8 years (64%).

Table 4.10: Prevalence of anaemia among the age groups

<table>
<thead>
<tr>
<th>Age group</th>
<th>Number of children N (%)</th>
<th>Anaemia n (%)</th>
<th>Mild n (%)</th>
<th>Moderate n (%)</th>
<th>Severe n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>All</td>
<td>120 (100.0)</td>
<td>118 (98.3)</td>
<td>7 (5.9)</td>
<td>61 (51.7)</td>
<td>50 (42.4)</td>
</tr>
<tr>
<td>&lt;5</td>
<td>44 (36.7)</td>
<td>44 (100.0)</td>
<td>5 (11.4)</td>
<td>25 (56.8)</td>
<td>14 (31.8)</td>
</tr>
<tr>
<td>5-8</td>
<td>61 (50.8)</td>
<td>59 (96.7)</td>
<td>1 (1.7)</td>
<td>20 (33.9)</td>
<td>38 (64.4)</td>
</tr>
<tr>
<td>9-12</td>
<td>15 (12.5)</td>
<td>15 (100)</td>
<td>-</td>
<td>7 (46.7)</td>
<td>8 (53.3)</td>
</tr>
</tbody>
</table>

(<5yrs; no anaemia-11.0g/dl or higher, mild-10-10.9g/dl, moderate-7-7.9g/dl and severe <7g/dl) (5-11yrs, no anaemia-11.5g/dl or higher, mild-11-11.4g/dl, moderate-8-10.9g/dl and severe <8g/dl) (WHO, 2011)

4.8 Nutrition education at the health centre

When caregivers were asked whether they receive nutrition education at the health centre, majority of them (74.2%) responded “Yes”. Participants reported receiving the nutrition education either from a doctor (62.8%) or a nurse (2.2%). They mentioned specific nutritional advice given to them to ensure the well-being of their children. Almost half (42.5%) of the caregivers said health care providers (doctor or nurse) advised them to give their children more fruits and vegetables. Others reported on giving their children foods that will make their bones strong (4.2%). This is presented in Table 4.11 below.
Table 4.11: Nutritional advice given to caregivers of children with sickle cell disease

<table>
<thead>
<tr>
<th>Nutritional advice</th>
<th>Frequency</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Give the child more fruits and vegetables</td>
<td>51</td>
<td>57.3</td>
</tr>
<tr>
<td>Give the child a lot of water and fruits</td>
<td>13</td>
<td>14.6</td>
</tr>
<tr>
<td>Give the child hot soup and more water</td>
<td>7</td>
<td>7.9</td>
</tr>
<tr>
<td>Provide the child with foods that will give more blood example ‘Kkontomire’ (local vegetable)</td>
<td>6</td>
<td>6.7</td>
</tr>
<tr>
<td>Give foods that will make the bones strong</td>
<td>5</td>
<td>5.6</td>
</tr>
<tr>
<td>Observe the child to eat well</td>
<td>4</td>
<td>4.5</td>
</tr>
<tr>
<td>Let the child eat little oil foods, more fruits and vegetables</td>
<td>3</td>
<td>3.4</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>89</strong></td>
<td><strong>74.2</strong></td>
</tr>
</tbody>
</table>

4.9 Factors associated with malnutrition

Table 4.12 shows the result of a multiple logistic regression run to determine factors that were associated with malnutrition among the study participants. From the model, only genotype could significantly predict stunting (p=0.01) after controlling for other variables. Children with SS genotype were 4.8 more likely to be stunted compared to SC genotype. In addition, children less than 5 years of age had a lower tendency (p = 0.08) to be underweight compared to children 9 - 12 years of age. Wasting and thinness were not included as dependent variables due to the small number of children within these categories.
Table 4.12: Multivariable model of factors associated with malnutrition

<table>
<thead>
<tr>
<th>Variable</th>
<th>Stunting (&lt;-2HAZ-score) OR (95% CI)</th>
<th>p-value</th>
<th>Underweight (&lt;-2WAZ-score) OR (95% CI)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>0.8 (0.3 - 1.8)</td>
<td>0.53</td>
<td>1.6 (0.6 - 4.4)</td>
<td>0.34</td>
</tr>
<tr>
<td>Female</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Age (years)</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt;5</td>
<td>0.7 (0.2 - 2.5)</td>
<td>0.55</td>
<td>0.2 (0.1 - 1.2)</td>
<td>0.08</td>
</tr>
<tr>
<td>5-8</td>
<td>0.5 (0.1 - 1.9)</td>
<td>0.31</td>
<td>0.4 (0.1 - 1.9)</td>
<td>0.25</td>
</tr>
<tr>
<td>9-12</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Age of Diagnosis</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>More than 1 year</td>
<td>1.7 (0.3 - 8.8)</td>
<td>0.55</td>
<td>4.3 (0.5 - 9.4)</td>
<td>0.19</td>
</tr>
<tr>
<td>&lt;1 year</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Genotype</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>SS</td>
<td>4.8 (1.4 - 16.1)</td>
<td><strong>0.01</strong></td>
<td>1.5 (0.4 - 4.9)</td>
<td>0.49</td>
</tr>
<tr>
<td>SC</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Nutrition Education</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>0.9 (0.3 - 2.7)</td>
<td>0.93</td>
<td>1.03 (0.3 - 3.4)</td>
<td>0.96</td>
</tr>
<tr>
<td>Yes</td>
<td>1</td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Past admission</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No</td>
<td>0.5 (0.2 - 1.2)</td>
<td>0.11</td>
<td>0.9 (0.4 - 2.8)</td>
<td>0.65</td>
</tr>
<tr>
<td>Yes</td>
<td>1</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Binary logistic regression adjusting for other variables in the model. Response variables: normal/stunted; normal/underweight; reference (stunted and underweight). **Significance at p<0.05.**

*Nutrition education to caregivers or parents; +Admission of the child in the past one year.
CHAPTER FIVE

5.0 DISCUSSION

5.1 Child and caregivers characteristics

Sickle cell disease is a chronic genetic disorder that affects most African descent. Findings from the study indicate that the disease is common in males compared to females. This is similar to the work of Koeta et al (2015) who reported more males than female children with sickle cell disease in a study to determine the presence of Cholelithiasis in sickle cell disease. Other authors, Mukherjee and Gangakhedkar (2004), Helvaci and Kaya, (2011), Deore and Zade (2014), Gray et al., (1992), Kamble and Chaturvedi (2000), Deshmukh and colleagues (2006) have documented similar findings. However, Ephraim and colleagues (2015) have reported otherwise.

Although most of the children were being catered for by both parents, a substantial number were catered for by single parents who were divorced. Sickle cell disease places not only a burden on affected individuals but families as well. Past studies have suggested that marital conflicts and divorce are common in families affected by SCD (Midence and Elander, 1994; Eddy and Walker, 1999).

Education provides people with knowledge to make informed decisions and reasonable choices. Moronkola and Fadairo (2006) have reported that more than half (63.6%) of their participants who had tertiary education knew their genotypes and all considered genetic counselling important before marriage. In this study, most of the caregivers had not attained at least secondary school (SHS) education. This could have led to them getting into marriage without genetic counselling although 5% of the caregivers with higher education (Tertiary level) had children with sickle cell disease. This demystifies the need to educate people on this chronic genetic disease to save affected children and reduce the burden associated with the disease.
5.2 Clinical information of participants

Homologous genotype, SS, often referred to as sickle cell anaemia is the most severe and common form of sickle cell disease (Hoban et al., 2015). We documented more children with SS genotype, than other genotypes (SC and SF). This is not different from some works carried out in Ghana (Adjei et al., 2015, Osei-Yeboah and Rodriques, 2011, Wilson et al., 2012, Ephraim et al., 2015) and India (Chawla et al., 2013).

It has been suggested that health care professionals treating children with SCD should be aware of nutrient deficiencies and educate families about ways to improve their children’s intakes of essential nutrients (Rovner et al., 2008). In this study, nutrition education was given to caregivers as part of medical care to improve on the quality of lives of the affected children. Caregivers reported some of the nutritional advice they received from the health centre including giving their children foods that would strengthen their bones and much fruits and vegetables. They were also advised to give much fluid. Although families are advised to give SCD children a lot of fluids to prevent erythrocyte dehydration, fluids should be taken between and after meals rather than just before meals to prevent appetite suppression (Kwachak et al., 2007). It is important to mention that the advice they mentioned were delivered to them by the medical doctor other than nutritionist/dieticians who are specially trained to provide medical nutrition therapy. Doctors may have less knowledge on issues relating to dietary management of chronic diseases. Additionally, the much workload on the health care system and the high patient to doctor ratio in Ghana, may affect the quality and depth of nutrition education given to caregivers with sickle cell children by doctors.

Newborn screening and early diagnosis of SCD is important to provide infants and children timely and better treatment. In Ghana and many other African countries, SCD diagnosis is often delayed and made after several visits to the hospital or clinic with acute illness, rather than early diagnosis by virtue of neonatal screening (Ohene-Frempong and Nkrumah, 1994). Our findings confirm such report. Despite the implementation of neonatal screening for SCD in Ghana, more
than half of the children recruited were diagnosed after a year in life while others were diagnosed as late as 7 years. Some parents/caregivers reported early diagnosis of their children but did not believe until the manifestation of the disease symptoms. Some emphasised that all their children were normal so their child cannot have the disease. A parent noted that even after diagnosis and bringing the child for medical attention, she still does not believe her child has the disease. This clearly shows the low level of knowledge of caregivers and parents of affected children on this chronic genetic disorder.

5.3 Dietary intake and nutritional status of participants

Nutritional studies of children with SCD have identified numerous deficits that likely contribute to poor nutritional status. In this study, dietary intake was not associated with nutritional status although a single 24-h recall of dietary intake is not an accurate assessment of nutrient intake for an individual. Our findings support earlier report by Zemel and colleagues (2007) who reported no relationship of dietary intakes with growth failure among US SCD children. Analysis of the 24 hour dietary recall showed that more than half of the children showed adequate intake for protein (95.8%), iron (59.2%), vitamin B12 (60.0%) and zinc (63.3%) but not for energy (24.2%). This is consistent with the findings of Gray et al (1992) who reported adequate intake of protein, iron and zinc compared to RDA yet the sickle cell children weighted less and were thin. This points to the fact that sickle cell children have increased nutrient requirements.

In this study, children were routinely prescribed 5 mg of folic acid. Children and adults with SCD are routinely prescribed folate supplements due to chronically increased haemolysis and haematopoiesis (Gray, et al., 1992, van der Dijs, et al., 1998). This present study shows low intake of folate; only 13.3% of the study children showed adequate intake of folate. This finding is consistent with two previous studies (Kennedy et al., 2001, Segal et al., 2004). In these studies, more than half of sickle cell children had inadequate intake of folate. The low folate intake support the basis for supplementation to compensate for the chronically increased haemolysis and haematopoiesis among patients with SCD.
In sickle cell patients, optimal intake of vitamin A may be important to improve iron absorption, enhance erythropoiesis and immunity, increase the mobilisation of iron from tissue stores and possibly affect red blood cell differentiation (Villamor et al., 2000). Less than half (40.8%) of the children had adequate intake of vitamin A. Schall et al., (2004) have reported similar finding. The low intake of vitamin A is in contrast to the findings of Finan et al., (1988) where sickle cell children dietary intake of vitamin A exceeded the RDA. In their study, the authors used a 3-day diet record which is not memory dependent and has been recognised as an accurate method of dietary assessment compared to the single 24 hour which is memory dependent and more prone to errors used in this study. This could have accounted for the differences in intake.

Protecting and maintaining red blood cell membranes from free radical damage is pertinent in the management of SCD. Experimentally, the antioxidant nutrients vitamin C and E have been shown to exhibit inhibitory role in red blood polymerisation and have been suggested that a cocktail of antioxidants would be effective in reducing the incidence and severity of sickle cell crisis (Puppalwar et al., 2015, Ohishi and Ohishi, 2001). Unfortunately, intakes of these antioxidant nutrients (vitamin C and E) were low among the study participants. This is not different from the work of Kwakwak et al., (2007) where sickle cell children had low intake of vitamin E and other nutrients. These low intakes may explain the high number of SCD related admissions (vaso-clusive crisis and anaemia) observed in this study.

Calcium is well-recognised for its role in strengthening the bones for erythropoiesis. This makes the mineral an important nutrient in sickle cell patients. Surprisingly, none of the children met the recommended requirement for calcium. This is in line with the findings of Meeuwes et al., (2013) where all Brazilian sickle cell children and young adults studied had a calcium intake below RDA. Other investigators (Arlet et al., 2013, Rovner et al., 2008, Lal et al., 2006, Buisson et al., 2004) have also reported lower intake of calcium among sickle cell patients. It is difficult to meet calcium requirements from other food sources without liberal consumption of dairy products (Shils and Shike, 2006). Although 38% and 18.3% of the children ate from milk and
milk products and dark green leafy vegetables food groups which are rich in calcium, they were not adequate to meet the recommended daily requirement. This could have accounted for the high number of vaso-occlusive related admissions observed in this study.

Kawchak et al (2007) evaluating the daily intake over the last 24 h in American children with SCD, showed a sub-optimal intake of many nutrients at all ages, including a low intake of vitamins D and E, calcium, folate, magnesium and zinc, declining in intake with increasing age, especially during adolescence. This current study confirms this finding; age trend showed that meeting nutrient requirement declined with age. At age 3, the children met RDA for six of the nutrients, this declined to four for children aged 4-8 years and three for children between 9 to 12 years. As age increases, dietary intake might have remained the same or may not be adequate to meet the increased nutrient requirements. This may explain the decline in meeting requirements for most of the nutrients with increase in age.

Malnutrition is common among children with sickle cell disease as indicated by delay in sexual maturation, deficits in weight and height, (Zemel et al., 2007) and low bone mineral density (Lal et al., 2006, Miller et al., 2006). Earlier study in Ghana reported high prevalence of malnutrition (61.3%) among sickle cell children attending routine check-up at Korle-Bu Teaching hospital (Osei-Yeboah et al., 2011). These authors reported significantly more stunted and underweight sickle cell children compared to their normal (HbAA) counterparts. This is similar to our findings, 38.0% of the participants were malnourished, 25.8% were stunted and 20.0% were underweight. The difference in prevalence may be due to the different age groups used (1 to 12 years versus 3-12 years). Children are particularly vulnerable to malnutrition in the first year of life especially when coupled with disease condition. Other possible reason could be due to the different medical treatments offered to affected children at these two different health facilities.

Sickle cell anaemia (SS) has been associated with stunting and wasting in a cohort of Tanzania children (Cox et al., 2011). This is consistent with our finding; SS genotype was significantly associated with stunting but not underweight. Undernutrition, increased nutrient requirements,
and low dietary intake often associated with the severe form of the disease (SS) may explain this finding.

5.4 Disease severity and Nutritional status

Disease severity is difficult to define for SCD due to the use of different criteria in assessment (Zemel et al., 2007). Severity of disease may affect dietary intake and influence nutritional status. In this study, SCD-related hospitalizations, average painful episodes and a measure of steady-state haemoglobin were used as markers of disease severity. Mandese et al., (2016) have recently documented that, inadequate nutritional intake, weight and BMI have a significant impact on SCD severity indices (haemoglobin level and number of days of hospitalization) in Canadian children with sickle cell disease. A similar finding has been reported by Zemel et al., (2007) where a significant association was observed between severity of disease and nutritional status among US children. In this study, there was no association between disease severity and nutritional status. This difference could be due to the variation in sample size (148 versus 120) and study design (longitudinal versus cross-sectional). The small sample size may have reduced the chance of providing a high statistical power to detect differences between the variables (disease severity and nutritional status).

Dietary diversity score used as an index of dietary intake in this study showed no significance with disease severity. This is in contrast with the findings of Mandese et al., (2016), who reported significant association of inadequate dietary intake with disease severity. In their study, severity of disease was assessed through number and days of hospital admissions and haemoglobin level. This difference can be explained by the method of disease severity classification and the study design (Observational versus cross-sectional). Other possible reason for this difference is the relation of dietary intake with single determinant of disease severity (either haemoglobin or number of hospital admissions).
5.5 Prevalence of anaemia

At birth, infants with SCD look normal and do not have anaemia, but with the synthesis of adult Hb, they develop chronic haemolytic anaemia that is present throughout life. From our results, haemoglobin level of the children varied from 5.3 to 11.8 g/dl with mean of 7.8 ±1.39 g/dl. This is in agreement with a study by Nikhar et al., (2012) who recorded a lower mean haemoglobin level among sickle cell patients from rural areas in India with a mean haemoglobin level of 7.8 ± 0.2 g/dl. Others (Buchowski et al., 2002; Daak et al., 2013; Cox et al., 2011) have also reported such a lower haemoglobin level among sickle cell patients.

Almost all the children (98.3%) were anaemic. The root cause of anaemia in sickle cell children is from chronic haemolysis due to loss of erythrocyte membrane integrity attributed to deficiency of antioxidant nutrients, folate or iron deficiency and depression of erythropoiesis (aplastic crisis) (Juwah et al, 2004). Folate is required for the production of red blood cells while vitamin C and E serve as potent inhibitors of sickle cell haemoglobin polymerisation (Pupalwar et al., 2015). The low intake of folate and antioxidants vitamins (E and C) by the children may have contributed to the high prevalence of anaemia observed in this study.

5.6 Strength and limitations of study

Strength of this study can be attributed to the fact that we extensively evaluated many micronutrients from the 24 hour dietary recall. To the best of our knowledge, this is the first study in Ghana on relationship of dietary intake, disease severity and nutritional status of sickle cell children. The small sample size is a cohort of children with SCD that were selected from children attending only one sickle cell clinic (PML) in Accra. We initially planned to increase our sample size by including sickle cell patients from Korle-Bu Teaching hospital. This was not done due to delay in obtaining ethical approval from the institutional review board (IRB) of Korle-Bu. The small sample size used in this study may not be a representative of the general population. Recall bias might have also influenced the process of recall of foods eaten during the past 7 days. Also, the use of the 24-hour recall is not representative of usual intake due to day-to-
day variation of food intake. This may have led to inaccurate estimation of children with lower intake of nutrients.
CHAPTER SIX

6.0 CONCLUSIONS AND RECOMMENDATIONS

6.1 CONCLUSION

This study shows that children with sickle cell disease have low intake of energy and micronutrients particularly calcium and the antioxidant nutrients, vitamin C and E. Age trend showed that meeting nutrient requirements declined with increase in age. The prevalence of malnutrition, a common feature among sickle cell children was 38%, with prevalence of stunting, underweight, thinness and wasting as 25.8%, 20.0%, 15.8%, and 6.8% respectively. Child’s genotype, (SS) significantly predicted stunting but not underweight.

There was no relationship between dietary intake, severity of illness, and nutritional status among the study children. Almost all the children (98.3%) were anaemic.

6.2 RECOMMENDATION

From the findings of the study, the following recommendations are being made:

- There is the need to develop comprehensive management coupling nutritional therapy to medical care to achieve the optimum goal of caring for affected children and improve nutritional status.

- Nutritional management should focus much on calcium-rich foods and antioxidants nutrients particularly vitamin C and E to reduce rapid erythrocyte haemolysis and chronic anaemia.

- Recommendation of fluid intake should be made in reference to between and after meals to avoid appetite suppression.

- Detailed nutrition education needs to be intensified for caregivers with children with sickle cell disease. This should be done by qualified nutritionists or dieticians at health centres. If possible, medical doctors should refer affected children/caregivers to these health practitioners after medical attention.
• There is the need to develop specific dietary recommendations much more than RDA for children with SCD due to increased nutrient demand associated with the disease process as it is done in conditions such as growth and pregnancy.

• Further studies could take into account detailed biochemical analyses to associate dietary intake with blood micronutrient status and also consider using a larger sample size possibly from different health facilities to generate more statistical power to make a robust inference about the population in question.
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University of Ghana http://ugspace.ug.edu.gh
APPENDIX I- INFORMED CONSENT FORM

TITLE: Dietary intake and nutritional status of children aged 3-12years with sickle cell disease.

Principal Investigator: Isaac Boadu

Supervisors
Dr. Agartha Ohemeng, Department of Nutrition and Food Science, University of Ghana.
Prof. Lorna Awo Renner, Department of Child Health, Korle-Bu Teaching Hospital.

General Information about Research
We are conducting a research study; the purpose is to assess the dietary intake and nutritional status of children (3-12yrs) with sickle cell disease to help develop a comprehensive national management plan for children with sickle cell disease. Your child is being invited to participate in this study because he/she has the sickle cell condition. If you agree to take part in this study, your child’s blood level will be measured by a prick on the finger by a well-trained biomedical Scientist. Small volume (drop) of blood (10µl) will be taken from the finger prick to determine his/her blood level. You can know your child’s blood level (haemoglobin) upon request. You will also be asked to provide information about the child’s condition/health, the food items that he/she took in the past 24 hours; in addition his/her height, weight and mid upper arm circumference will be measured. The interview will take approximately 30 minutes of your time.

Possible Risks and Discomforts
There is no risk involved in taking part in this study. Your child may experience little discomfort from the finger prick. This will be minimised by the research team by ensuring the necessary precautions.

Possible Benefits
The study provides no direct benefit to you or your child. It is expected that the findings will help us understand the dietary intake and nutritional status of the children to help develop a comprehensive national management plan and also advance scientific knowledge.
Confidentiality

Information provided will be kept confidential and protected to the best of our ability. When the results of this research are published or discussed in any conference, no information will be included that will reveal your child’s identity. Only investigators involved in this study may sometimes look at his/her records. The records of the study will be kept at the University of Ghana.

Compensation

At the end of the study, a drink (Kalyppo) and biscuit will be given to your child in appreciation for his/her participation.

Voluntary Participation and Right to Leave the Research

Your child’s participation is voluntary. You may decide to stop your child’s participation at any time without a penalty. You have the right to ask that any data supplied to that point be withdrawn/destroyed. We will ask your child’s permission before we begin the study, and they can stop at any point.

Contacts for Additional Information

You are encouraged to ask any questions at any time of the study. For answers to any pertinent questions about the research, you may contact the principal investigator, Isaac Boadu (telephone number: 0207248047 or 0243881993. E-mail: domainjnr14@gmail.com) or the supervisor, Dr. Ohemeng (telephone number: 0244862606. E-mail: anohemeng@ug.edu.gh).

Your rights as a Participant

This research has been reviewed and approved by the Ethical Review Committee of the Ghana Health Service (GHS-ERC) and the institutional review board of the college of Basic and applied science (ECBAS) at the University of Ghana. If you have any questions about your rights as a research participant you can contact the administrator of GHS-ERC, Hannah Frimpong through office line: +233 302681109; mobile: +233(0) 243 235225 or 0507041223; or email: Hannah.Frimpong@ghsmail.org
B. PARTICIPANT STATEMENT AND SIGNATURE

By signing below, you are agreeing that:

(1) You have read or someone has read to you and understood the participant information sheet.

(2) Questions about your child’s participation in this study have been answered satisfactorily.

(3) You are willing for your child to take part in this research study voluntarily.

________________  __________  ______________________
Name               Date                        Signature or thumbprint of Participant

(Thumbprint for those who cannot read and write)

If volunteers cannot read the form themselves, a witness must sign here:

I was present while the benefits, risks and procedures were read to the volunteer. All questions were answered and the volunteer has agreed to take part in the research.

________________  __________  ______________________
Name               Date                        Signature of witness

C. INVESTIGATOR STATEMENT AND SIGNATURE

I certify that the participant has been given ample time to read and have understood the study.

All questions and clarifications raised by the participant have been addressed.

________________  __________  ______________________
Name               Date                        Signature of Person Who Obtained Consent
APPENDIX II-QUESTIONNAIRE
DEPARTMENT OF NUTRITION AND FOOD SCIENCE
UNIVERSITY OF GHANA, LEGON PROJECT

TITLE:
Dietary intake and Nutritional status of children aged 3-12years with sickle cell disease.

SECTION A
Socio-demographic characteristics

Please take available information from their medical records
(Please indicate in the brackets the code that bears the response)

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<tbody>
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<td>A1.</td>
<td>Age......................................... (Years)</td>
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<td>A2.</td>
<td>Sex (1=male 2=female) [ ]</td>
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<td>A3.</td>
<td>Place of residence..............................................................</td>
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<td>A4.</td>
<td>Educational level (0 = No formal education, 1= Preschool, 2=Primary 3=JHS [ ]</td>
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<td>A5.</td>
<td>Ethnicity (1= Ga 2=Ewe 3=Akan 4= other (specify))...................... [ ]</td>
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<td>A6.</td>
<td>Religion (1= Christian 2= Moslem 3= traditionalist 4= other (specify))...... [ ]</td>
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<td>A7.</td>
<td>Nationality (1 = Ghanaian 2=other (specify))................................. [ ]</td>
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<td>A8.</td>
<td>Guardian 1= both parents 2 = Single parent 3= Grandmother 4=other (specify)............ [ ]</td>
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<td>A9.</td>
<td>Educational level of main provider 1= no formal education 2= Primary 3 = JHS 4 = SHS 4= tertiary [ ]</td>
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<td>A10.</td>
<td>Occupation of main provider [ ]</td>
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<td>A11.</td>
<td>Marital status of main provider 1=Married 2=Single 3= Widowed/Divorced [ ]</td>
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</table>
SECTION B
Clinical information

Please take available information from their medical records

B11. Are you suffering from other diseases apart from sickle cell disease? [ ]
   1=Yes  2=No

B12. What type of disease? .................................................................

B13. Type of sickle cell disease [ ]
   1=SS  2=SC  3=S/B⁺  4=S/B⁻

B14. At what age were you diagnosed of the disease? [ ]
   1=at birth  2=1year  3=2 years  4=3 years  5=other (specify)..............

B15. At what age did you start attending the sickle cell clinic? [ ]
   1=1year  2=2 years  3=3 years  4=4 years  5=other (specify)..............

B16. If not same as diagnostic age, ask why?

B17. Do you experience crises? [ ]
   1=Yes  2=No

B18. How often do you experience the crisis? [ ]
   1=very often  2=sometimes  3=other (specify).............................

B19. Average painful crises per year.................................

B20. What do you do when you have the crisis? [ ]
   1=take medication  2=eat/take fluids  3=nothing,  4=other (specify)........

B21. If you take medication what medication do you take? [ ]
   1=folic acid  2=aspirin  3=Paracetamol  4=other (specify)................

B22. Do you give the child certain foods during crisis? [ ]
   1=Yes  2=No

B23. If yes, what kind of foods? ..........................................................

B24. Do you receive nutrition education at the health centre? [ ]
   1=Yes  2=No

B25. If yes, from who?
   1=nurse  2=Doctor  3=Nutritionist/Dietician

B26. What nutritional advice do you receive?

B27. Have you been on admission in the past 12 months? 1=Yes  2=No [ ]
B28. Reason for admission [  ]
1=Bone pain  2=abdominal pain  3=Chest pain  4=Malaria  5=anaemia
6=other......

B29. Number of admissions in the past 12 months? ..............................................

B30. How long were you on admission? [  ]
1= 3 days  2= one week 3= one month  4= other (specify)............................

B31. Have you received blood transfusion in the past 12 months? [  ]
1=Yes  2= No

B32. How many times have you been transfused? ..................................................

B33. Amount of blood transfused in the past 12 months ........................................

B34. How often do you visit the sickle cell clinic? [  ]
1= weekly  2= two weeks  3=monthly  4= when I have crisis
5= other (specify)..........................................................

B35. Do you miss clinic appointment?
1=Yes  2= No [  ]

B36. If yes why........................................................................................................

B37. What is your opinion on the treatment that you receive at this hospital?
..........................................................................................................................

..........................................................................................................................
SECTION C- Dietary Assessment

C1.  24-hr Dietary recall

Kindly tell me all the foods and drinks you ate from the past 24- hours up to now

<table>
<thead>
<tr>
<th>Time</th>
<th>Details of food and drink</th>
<th>Estimated amount eaten</th>
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</table>
C2. **Food group frequency (within 24 hours and 7 days)**

Ask whether the child has eaten each of the food items in the last 24 hours and the past 7 days and how many days in the past 7-days the child has eaten it.

<table>
<thead>
<tr>
<th>FOOD GROUP</th>
<th>Has the child eating this in the last 24 hours</th>
<th>Has child eating this in the last seven days?</th>
<th>How many days in the last 7-day did your child eat it?</th>
<th>Number of days</th>
</tr>
</thead>
<tbody>
<tr>
<td>STARCHY STAPLES (cassava, yam, bread, porridge, corn/maize, rice, wheat, sorghum, millet, potatoes)</td>
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<td>DARK GREEN LEAFY VEGETABLES AND RED PALM OIL <em>(kontonmire, ayoyo, cassava leaves)</em></td>
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<td>VITAMIN A RICH FRUITS AND VEGETABLES (carrot, ripe mango, ripe papaya)</td>
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<td>OTHER FRUITS AND VEGETABLES (Garden eggs, tomato, onion, other fruits, including wild fruits and 100% fruit juice made from these)</td>
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<td>ORGAN MEAT (liver, kidney, heart or other organ meats or blood-based foods)</td>
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<td>MEAT AND FISH (beef, pork, lamb, goat, rabbit, game, chicken, duck, other birds, fresh or dried fish or shellfish)</td>
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<tr>
<td>EGGS (Eggs from chicken, duck, guinea fowl or any other egg)</td>
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<tr>
<td>LEGUMES, NUTS AND SEEDS dried beans, dried peas, groundnut, seeds or foods made from these (e.g. peanut butter)</td>
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<tr>
<td>MILK AND MILK PRODUCTS (Milk, cheese, yogurt or other milk Products)</td>
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SECTION D

D1. Anthropometry

Kindly measure the child’s weight, height and MUAC twice and record

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<th>1ST</th>
<th>2ND</th>
<th>Average</th>
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<td>Weight (Kg)</td>
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<tr>
<td>Height (cm)</td>
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<tr>
<td>Muac (cm)</td>
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D2. Haemoglobin level (g/dl)……………………………………