PREVALENCE OF SICKLE CELL DISEASE AMONG CHILDREN UNDER 10 YEARS ADMITTED AT MBALE REGIONAL REFERRAL HOSPITAL BETWEEN MAY 2014 AND JULY 2014.

BY

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(BMS/0050/91/DU)

A RESEARCH PROJECT REPORT SUBMITTED IN PARTIAL FULFILLMENT FOR THE AWARD OF BACHERLORS DEGREE IN MEDICINE AND SURGERY AT KAMPALA INTERNTIONAL UNIVERSITY WESTERN CAMPUS

JULY 2014-OCTOBER 2014

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DECLARATION

To the best of my knowledge and belief the work here is authentic, original and has never been submitted anywhere either partially or in full for the award of any Degree/Diploma/Certificate in any university.

I also declare that the use of literature from other sources has been accredited to their respective authors.

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SUPERVISOR'S APPROVAL.

This report has been approved for submission to the faculty of clinical medicine and dentistry. Kampala International University in partial fulfilment for the award of Bachelor of Medicine and Surgery at Kampala International University:

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DEDICATION

I dedicate this research to my family, especially my father and mother Mr. and Mrs. Masaba Michael for your unconditional support with my studies. I am honored to have you as my parents. Thank you for believing in me and giving me a chance to prove and improve myself through all my walks of life please do not ever change. I love you.

To my siblings Martha, Davis, Doreen, Sadiq and Laban. Hoping that with this research I have proven to you that there is no mountain higher as long as God is on our side. Hoping that you will always be able to standout and fulfil your dreams

ACKNOLWEDGMENT

I would like to thank the Almighty God for giving me the opportunity to be where I am, what I've become and what he has done and so far accomplished in my life, I would also like to acknowledge my supervisor professor EDUARDO CODISPOTI whose scholarly advice, help, constant encouragement and support have contributed generously to my study. My gratitude goes to medical library staff for making reference books available for my study.

My appreciation also goes to Mbale Regional Referral Hospital director, the paediatric clinic staff, and the staff at the records who willingly accepted me and helped me in carrying out the study.

To you all, may the good Lord Almighty reward you accordingly?

ABSTRACT

Sickle cell disease is a major public health problem in the Sub-Saharan Africa contributing significantly to morbidity and mortality in children under ten years. SCA accounts for approximately 16.2% of all paediatric deaths in Uganda. The pattern of SCD inheritance, however, predicts likely changes in the prevalence and distribution of the SCD. The objective of the study therefore was to establish the current prevalence oof sickle cell disease as well as associated factors among children under ten years admitted to Mbale Regional Referral Hospital pediatric unit.

The study was descriptive and cross sectional in which both qualitative and quantitative methods were used for data collection.

The ward admission book was used as a basis to capture those admitted and fit into the specified age than ten years.

In addition questionnaire was admitted to the ward stuff and to the in charge to obtain a little more data to find out how the children presented before being admitted to the hospital, any other information relevant to the study including common disease and any preventative services the centre offers to the community around.

Data was analysed using Microsoft excel, discussed and conclusions made to the findings of the study. The dissertation will thereafter be submitted to the KIU examination board.

A total number of 6790 patients records were analysed, most of them, 53% being female. Majority of the patients were Bagisu followed by the Bagwere and a small number of the other tribes.

The study showed that the prevalence of SCD (Haemoglobin ss) was 2.2% being higher than the 2010 studies which showed a prevalence of 1.7%.

SCD exists in Mbale Regional Referral Hospital in quite significant numbers to warrant the ministry of health and the district health service team’s action.

LIST OF ABBREVIATIONS

%	Percentage
DHMT	District Health Management Team
DNA	Deoxyribonucleic Acid
g/dl	Grams per decilitre
Hb	Haemoglobin
HbAs	Sickle cell trait
Hbss	Sickle cell anaemia.
ID	Identification
K.I.U	Kampala international university
MCH	Maternal and Child Health
MOH	Ministry of Health
Qty	Quantity
SCD	Sickle cell disease
SCT	Sickle cell trait
SPSS	Statistical Package for the Social Sciences
UDHS	Uganda demographic health survey
UHMIS	Uganda health management information system
USA	United States of America
WHO	World Health Organization

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CHAPTER ONE

1. INTRODUCTION

1.1 Background Information

Sickle cell disease (SCD) is a genetic disorder, and the sickle cell anemia (HbSS) is the more severe genotype. The disease is characterized by the presence of the hemoglobin S (HbS), where valine replaces glutamic acid at the beta globin chain that has a single point mutation at the sixth codon of the b-globin (HBB) gene.

Sickle cell disease clinical outcome vary widely from mild to severe and has been associated with multi-organ damage and risk of early mortality with acute and chronic clinical manifestations, including vaso-occlusive episodes, painful crisis, tissue ischemia/reperfusion injury, haemolysis, impaired blood flow as a result of intravascular sickling in capillary and vessels, inflammation processes and high susceptibility to infection, encephalic vascular accident (EVA), dactylitis, leg ulceration, pulmonary hypertension, and acute chest syndrome.

About 5% of the world's population carry genes responsible for haemoglobinopathies. Nearly each year about 300,000 infants are born with major haemoglobin disorders. Globally there are more carriers than (healthy people who have inherited only one mutant gene from one parent) of thalassemia than of sickle cell anaemia, but the high frequency of the sickle cell gene in certain areas leads to a high rate of the affected new-borns (WHO 2006)

Sickle cell anaemia is particularly high among people whose ancestors come from sub-Saharan Africa, India, and Saudi Arabia and Mediterranean countries. Migration raised the frequency of the gene in American continent. In some areas of sub-Sahara Africa, up to 2% of the children are born with SCD. The prevalence of sickle cell trait (healthy carries who have inherited the mutant gene from only one parent) ranges from 10% and 40% across equatorial Africa and decreases to between 1% and 2% on the north African coast and <1% in south Africa. (WHO 2006)

The impact of sickle cell disease on health in same areas as measured by under 10 mortality indicates that it contributes the equivalent of 4% of under-ten death on the African continent. More than 9% of these death occur in West Africa more importantly Nigeria and Ghana (Rhim MC Ganbo A 2003).

In the United States of America, the median survival as estimated in 1994 was 42 years for men and 48 years for women, however there is no firm data on the survival of patients with sickle cell anaemia in Uganda and Africa as whole. SCA accounts for about 16% of all paediatric deaths in Uganda. (okuil-etal 2010).

In Sub-Sahara Africa mortality is expected to be much higher than in many areas, estimates derived from the age structure of the populations attending clinics, suggest that more than half of those with sickle cell anaemia have died by the age of 10 years usually from infections including malaria, pneumococcal sepsis and from anaemia. (WHO 2006)

The first survey on sickle cell disease (SCD) done in Uganda in 1949, reported the district of Bundibugyo in Western Uganda to have the highest sickle cell trait (SCT) prevalence (45%). This is believed to be the highest in the whole world. According to the same survey, the prevalence of SCT in the districts of Mbale and Sironko in the East was 20-28%, whilst the districts of Mbarara and Ntungamo in the West had 1-5%. No follow-up surveys have been conducted over the past 60 years. SCA accounts for approximately 16.2% of all paediatric deaths in Uganda. The pattern of SCT inheritance, however, predicts likely changes in the prevalence and distribution of the SCT. The objective of the study therefore is to establish the current prevalence of the SCT in Mbale.

1.2 Statement of the problem.

About twenty million people world-wide have sickle cell disease and nearly 15million (70% of these are in Africa (Aliyu &Gregory J.Kato, 2008) some studies done indicate that it's the most affected with 200,000 new borns affected by SCA per year with the male to female ratio of 2:1. (Diallo D Tcernia 2003). Reports from Ghana, indicate that 15,000 children are born with sickle cell disease (SCD) annually (Kowaku O 2010). In Benin the sickle cell trait (SCT) prevalence is estimated to be 25% (Rhim MC Ganbo A 2003), While in Nigeria it ranges from 24-25% (Seargent GR 2005).

The prevalence of sickle cell gene among Chagaa tribe in Tanzania is about 4%. In all these African countries, the concentration of the SCT has been found to be highest in specified sub-populations likely due to tribal conservative marriages (Akannjaun O 1994).

The autosomal recessive pattern of inheritance assumed SCT however, predicts the changes in the population-wide dynamics, ease of movement and inter-tribal marriages which would change the SCT distribution with in these communities (Cvallin Sefroza et al).

Sickle cell accounts for a significant number of morbidity and mortality. It's the 3rd leading cause of child hospitalisation after malaria and pneumonia in most parts of sub-Saharan Africa (Dialo and Tchernia 2003).

Data available at the MOH Uganda on sickle cell disease (SCD) was done in 1949, reported the district of Bundibugyo in Western Uganda to have the highest sickle cell trait (SCT) prevalence (45%). This is believed to be the highest in the whole world. According to the same survey, the prevalence of SCT in the districts of Mbale and Sironko in the East was 20-28%. (Lenham H 1949)

Other studies done show that about 900,000 children are born in Uganda annually and of these approximately 2.8% have sickle cell anaemia and about 20,000 (70-80%) of these children born with SCA presumably die before their 5th birth day. This number contributes 16.2% of all children who die annually in Uganda. (UDHS 2006).

Data from the HMIS and laboratory in Mbale indicates that large numbers of children less than 10 years of age have been transfused due to malaria and anaemia. As sickle cell and anaemia are closely related this study tries to find out the prevalence of SCD at the facility, among children under five that have been admitted to the hospital in this period of May 2014 to July 2014.

1.3.1 General objective

To establish the prevalence of sickle cell disease among children who are admitted to the paediatric ward for the period between May 2014 and July 2014.

1.3.2 Specific objectives

1. To determine the number of sickle cell disease children among those admitted on the paediatric ward for this period of time.
2. To assess the distribution of SCD among the different ethnic groups.
3. To establish the levels of haemoglobin with which sickle cell children present with.

1.4 Justification of the study.

SCD is one of the leading causes of morbidity and mortality among children under ten years in the hospital, it's important at this point in time to carry out a study to determine the prevalence of the disease.

The study will also provide information to relevant authorities, policy makers and organizations that might assist in the prevention and control of sickle cell disease .The findings of the study would also act as a basis of reference for further research and creating opportunities for health education and promotion in the management and control of the disease.

It's also a requirement for the award of a bachelor of medicine and bachelor of surgery degree at Kampala international university.

Sickle cell is a neglected disease in Uganda as evidenced from the fact that it does not form part of the Uganda child health priorities for the ministry of health despite high deaths in the country, so it's important that new studies are done to awaken program planners for example the Uganda sickle cell association and the ministry of health. There is need to bridge this knowledge gap, get current, prevalence and possibly draw stakeholders attention to the health problem in the district that is not being looked at keenly.

1.5 Hypothesis

There is high prevalence of anaemia among children with SCD.

There is a high prevalence malaria among children with SCD.

1.6 Importance of the study

The study addressed health, socio-economic and environmental issues related to sickle cell disease among children in Mbale district. The information obtained from this study will be used by government and the non-governmental organizations to design appropriate interventions and make policies that will reduce the morbidity and mortality of children due to sickle cell disease.

1.7 Scope of the study

The study focused on determining the prevalence of SCD Among children bellow 10 years in Mbale District. The study addressed health, socio-economic and environmental issues related to sickle cell disease in Mbale District.

1.7.1 Geographical scope

Mbale District is bordered by Sironko District to the north, Bududa District to the northeast, Manafwa District to the southeast, Tororo District to the south, Butaleja District to the southwest and Budaka District to the west. Pallisa District and Kumi District lie to the northwest of Mbale District. Mbale, the largest town in the district and the location of the district headquarters, is located approximately 245 kilometres by road, northeast of Kampala, the capital of Uganda, and the largest city in that country. The coordinates of the district are: 00 57N, 34 20E. It has an area of 518.8 square kilometres (200.3 sq. mi). The population of Mbale District is estimated at 441,300 with an annual population growth rate of 2.5%.

1.7.2 Content scope

The project determined the prevalence of SCD, consequences of poor health care and illnesses common too SCD children hence identify gaps in preventing and managing sickle cell related conditions among children in Mbale District.

1.7.3 Time scope

The research process started with topic selection in July 2014 followed by proposal write up and it’s due to end with a research report submission in October 2014.

CHAPTER TWO

2. LITERATURE REVIEW

2.1 Definition of sickle-cell disease.

Sickle-cell disease (SCD), or sickle-cell anaemia (SCA) is a hereditary blood disorder, characterized by an abnormality in the oxygen-carrying haemoglobin molecule in red blood cells that leads to a propensity for the cells to assume an abnormal, rigid, sickle-like shape under certain circumstances. Sickle-cell disease is associated with a number of acute and chronic health problems, such as severe infections and attacks of severe pain ("sickle-cell crisis"), and an increased risk of death. Sickle-cell disease occurs when a person inherits two abnormal copies of the haemoglobin gene, one from each parent. Several subtypes exist, depending on the exact mutation in each haemoglobin gene. A person with a single abnormal copy does not experience symptoms and is said to have sickle cell trait.

The inheritance of sickle cell anaemia occurs via an autosomal recessive gene with both parents, in general, asymptomatic carriers of a single affected gene (heterozygous), transmitting the defective gene to their child, who therefore is homozygous (Hb SS).

Clinical manifestations are observed only in homozygous individuals for Hb S (Hb SS), resulting in sickle cell anaemia.

2.2 Epidemiology

In studies that have been done worldwide it has been observed that sickle cell disease (SCD) affects millions of people throughout the world and in particular common among those whose ancestor came from the sub-Saharan Africa; Spanish speaking regions in the Western hemisphere (south America, the Caribbean and central America); Saudi Arabia, India; and Mediterranean countries such as turkey, Greece and Italy (world health report 2004).

Sickle cell disease occur more commonly among people from parts of the world were malaria is or was common. It’s believed that people who carry the sickle cell trait are less likely to have severe forms of malaria. (WHO report 2004). The highest frequency of sickle cell disease is found in the Tropics, particularly in the Sub Saharan Africa, India and the middle-East. (Weatherall DJ,Cleg JB 2001). Migration of substantial populations from these high prevalence areas to low prevalence areas like Europe has drastically increased in recent decades and in same European countries sickle cell disease has now overtaken familiar

genetic conditions such as haemophilia and cystic fibrosis. (Roberts I Demontalemdart July 2007)

Studies in Africa indicate that three quarters of sickle cell cases occur in Africa. A recent WHO study report estimated that around 2% of new born in Nigeria were affected by sickle cell anaemia, giving a total of 150,000 affected children born every year in Nigeria alone. The carrier frequency ranges between 10% and 40% across equatorial Africa, 1-2% in North Africa and less than 1% in South Africa (WHO 2006)

In the USA the prevalence of the disease is approximately 1 in 5,000, mostly affecting Americans of Sub Saharan African descent, according to the National institute of health. (National lung and heart institute). In the about 1 out of 500 African- Americans children and 1:36,000 Hispanic Americans born will have sickle cell anaemia. It's estimated that sickle cell Disease (SCD) affects 90,000 Americans.

2.3 Pathophysiology of sickle cell disease

The disease occurs due to a mutation of the beta globin gene of haemoglobin, causing a substitution of the glutamic amino acid for valine at position 6 of the beta chain, thereby producing an abnormal haemoglobin, called haemoglobin S (Hb S), instead of normal haemoglobin, haemoglobin A (Hb A). With modified physicochemical characteristics, the molecules of haemoglobin S suffer polymerization and precipitation, leading to a change in form, a deformity of red blood cells which become sickle-shaped. In this case, the viscosity of the blood increases due to the formation of tactoids.

During foetal and early postnatal life, the lack of expression of the Hb SS phenotype is explained by the production of foetal haemoglobin (Hb F) which is sufficient to limit, by dilution, the effects of sickling. As the red cells that emerge from the bone marrow carry increasing amounts of Hb S and smaller amounts of Hb F, the results of sickling gradually appear. Therefore, new-borns begin to manifest the disease from the sixth month of life, when the amount of Hb F begins to approach adult levels.

The clinical manifestations of sickle cell disease are the result of two characteristic processes: severe anaemia and vaso-occlusion. Anaemia results from a shorter half-life of the red blood cells containing primarily Hb S. While normal red blood cells circulate for about 120 days, those containing Hb S only last for 10 to 20 days, leading patients to present moderate to severe anaemia. The second process, which is physiopathologically more complicated, is

vaso-occlusion. The intravascular effect from the spatial change of haemoglobin leads to the formation of helical bundles that greatly alter the membrane's permeability to ions, and the red cell/blood vessel and the red cell aggregation/red cell ratios.

The effects of the phenomena of vaso-occlusion vary in intensity but include tissue ischemia, painful episodes, acute osteoid-articular or abdominal crises and chronic organic injuries such as functional asplenia, cerebral-vascular disease and kidney, heart and lung failure; patients require frequent hospitalization.

Although the sickle cell trait (Hb AS) is usually asymptomatic, there are reports of sudden death and medical complications such as haematuria, hyposthenia, pulmonary embolism and splenic infarction, especially when carriers are exposed to extreme conditions of low oxygen tension, such as in strenuous physical exertion, depressurization of a flight cabin, and high altitude environments.

In children, infections caused by encapsulated bacteria and intra-splenic vaso-occlusion (splenic sequestration) are the main causes of mortality. These begin after the first two to three months of life and affect 20-25% of children in the first 5 years of age. Children who have overcome this initial barrier, face the effects of chronic vaso-occlusion. Over the years, these small strokes are the determining factor for the impairment of organs, leading to pulmonary, liver or brain infarction, kidney failure and retardation in growth and sexual maturation, with progressive impairment of multiple organs. These phenomena significantly reduce the quality of life of individuals, increase need for medical care, and diminish the capacity to work and life expectancy.

The benefits of early diagnosis and intervention in the monitoring of sickle cell disease have led to widespread use of education programs to detect these conditions. Through neonatal screening programs, it is possible to reduce morbidity and mortality in the first five years of life. Furthermore, the prophylactic use of penicillin, the administration of pneumococcal vaccine and intensive care significantly increase the survival and quality of life of patients with sickle cell disease, reducing and extenuating the consequences of clinical complications.

2.4 Clinical features of sickle cell anaemia.

Exacerbations of anaemia may be associated with haemolytic crises, acute splenic sequestration (though this is probably less common in Africa than in the West Indies) and acute bone marrow aplasia, often due to parvovirus B19. In addition to anaemia and jaundice,

individuals with untreated HbSS are often stunted and may have deformities of the facial bones such a frontal bossing due to bone marrow expansion.

Growth and development, including puberty, are delayed and education and employment opportunities may be impaired due to repeated bouts of ill health.

Tissue infarction leads to swelling, severe pain and fever. In children under 2 years, infarcts occur typically in the small bones of the hands and feet (Fig. 67.2). In older children and adults, long bones are affected. Degenerative changes are common as a late consequence of infarction and infection and include avascular necrosis of bones, chronic leg ulcers, duodenal ulcers, retinopathy and progressive bone marrow failure.

Individuals with HbSS have an increased risk of infection because of defective complement activation, hyposplenism and diminished cell-mediated immunity. Strep. pneumoniae and Salmonella spp. are the most common bacterial organisms (Williams et al., 2009).

Acute viral infections are particularly severe, and may precipitate infarctive crises. Acute chest syndrome occurs in approximately 30 per cent of sickle cell patients and is characterized by a life-threatening deterioration in respiratory function and arterial oxygen tension.

Any organ can be damaged in sickle cell disease including the heart, lungs, eyes, and genitourinary and hepatobiliary systems.

Neurological complications, particularly strokes, are one of the most devastating consequences. Although many patients recover reasonable function after a stroke, permanent loss of neuropsychometric performance is common.

2.5 Diagnosis

The diagnosis of SCD is suggested by the family history, and the characteristic clinical features and peripheral blood picture (sickle cells, target cells, macrocytes, polychromasia, nucleated red cells and

Howell–Jolly bodies) and a positive HbS solubility test. Confirmation is by haemoglobin electrophoresis, isoelectric focusing or high performance liquid chromatography.

CHAPTER THREE

3 RESEARCH METHODOLOGY

3.1 Study Area.

The study area was Mbale Regional Referral Hospital and the study was among patients drawn from districts around the hospital, but not limited to since same patients are referrals from as far as Karamoja and even Busia. The hospital was purposely selected because it always attracts a great number of paediatric patients possibly because it's a referral centre with facilities to handle a great deal of SCD complications. And because of its fairly constant turn up provides a good sampling area.

3.2 study Population

The study population included all the children who were admitted to Mbale Regional Referral Hospital paediatric ward during the three month period as from May to July 2014.

3.3 Study Design

The study was a cross-sectional descriptive study where all children under ten years admitted at Mbale Regional Referral Hospital Paediatric ward were recruited and questionnaires administered after signing informed consent forms. Both qualitative and quantitative data collection methods were used.

3.2 Sample size and selection of eligible respondents

3.3.1 Sampling Technique

The survey was in form a census were all children less than 10 ten years admitted to Mbale Regional Referral Hospital paediatric ward were sampled.

3.3.3 Exclusion Criteria

Any child above the age of ten years was automatically excluded from the study, and further still excluded from the study were those children who according to the opinion do not qualify to be admitted to paediatric word.

3.3.4 Inclusion Criteria

Any child under the age of ten who was admitted to the paediatric word as judged by the clinician responsible, qualified to be included in the study.

3.3.5 Selection of quantitative data:

All children bellow under 10 admitted to the paediatric ward in this period from May 2014 to July 2014 were included in the sample.

3.3.6 Selection of qualitative data:

Record review was be done for all paediatric cases who had been tested for sickle cell disease for the last year 2013. Scheduled group discussions with the paediatrics health care workers and also scheduled interview with the paediatric ward in charge was carried out.

3.4 Data Collection Procedure

Data was collected from register book of the paediatric ward. And the laboratory and this ensured that the children diagnosed for SCD had their Hb estimated in the laboratory.

3.4.1 Data collection instruments.

The ward record book, lab register and a specifically prepared checklist, Questionnaire, stationary were used to capture data.

3.5 Data Analysis

Data was processed and analyzed using MS WORD and EXCEL Microsoft programme. It's presented by use of tables, pie charts and histograms for easy interpretation and analysis.

3.6 Ethical Considerations

The study was be approved by Mbale District Health Management Team and informed consent was obtained from all the participants. All procedures that followed were in accordance with the ethical standards of the Ministry of Health (MOH). Ethical approval was obtained from Kampala International University Ethics Committee before carrying out this research project. All the information given by the participants will be confidential. The identity of the participants will not be revealed.

3.7 Limitations of the study

Mbale regional referral Hospital is a referral centre and therefore attracts many clients from far because of its superiority in providing specialized service this also may in a way influence the patient choice in seeking health care more so for those with SCD. These could introduce some biases in the study and may limit its scope.

Though not many and not well established, there are some public and private clinics that provide care for children with SCD. This may cause same children to attend those clinics.

3.8 Minimization of the biases

To reduce bias related to the sampling procedure, the study only focused on children who seek paediatric health care at Mbale Regional Referral Hospital. Though there are other health facilities that provide similar services, assumption is made that the children were consistent in their health seeking behaviour. Again, because most clients from within Mbale municipality go to this hospital, it is expected that they fairly represent Mbale's population. This also reduces the influences introduced by referrals from outside. To reduce biases during the data collection period, the enumerators were trained on how to conduct interview and collect information. The data collection tools were pre-tested in different but similar areas. The principal researcher supervised the enumerators and verified the data collected on daily basis.

3.9 Plans for dissemination of data.

Copies of the research report are submitted to the faculty of clinical medicine and dentistry of Kampala International University, the main library of KIU western campus, and the Health management team of Mbale District.

CHAPTER FOUR: STUDY FINDINGS

In this study, records of 6790 children under 10 years admitted to the paediatric unit in a period of three months between May 2014 and July 2014 at Mbale Regional Referral Hospital.

4.1. QUANTITATIVE RESULTS.

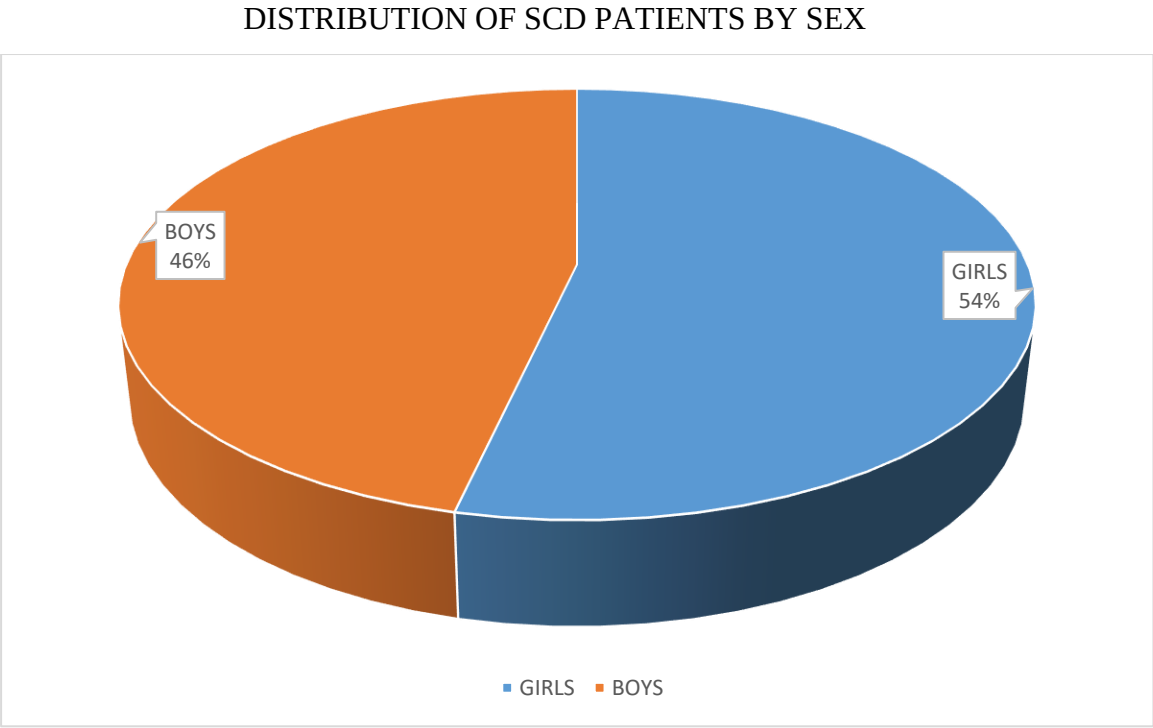
4.1.1. Demographic data

Table 1. Proportion of SCD in the general admissions.

Total no of admissions bellow 10	SDC Positives	percentage
6790	146	2.2%

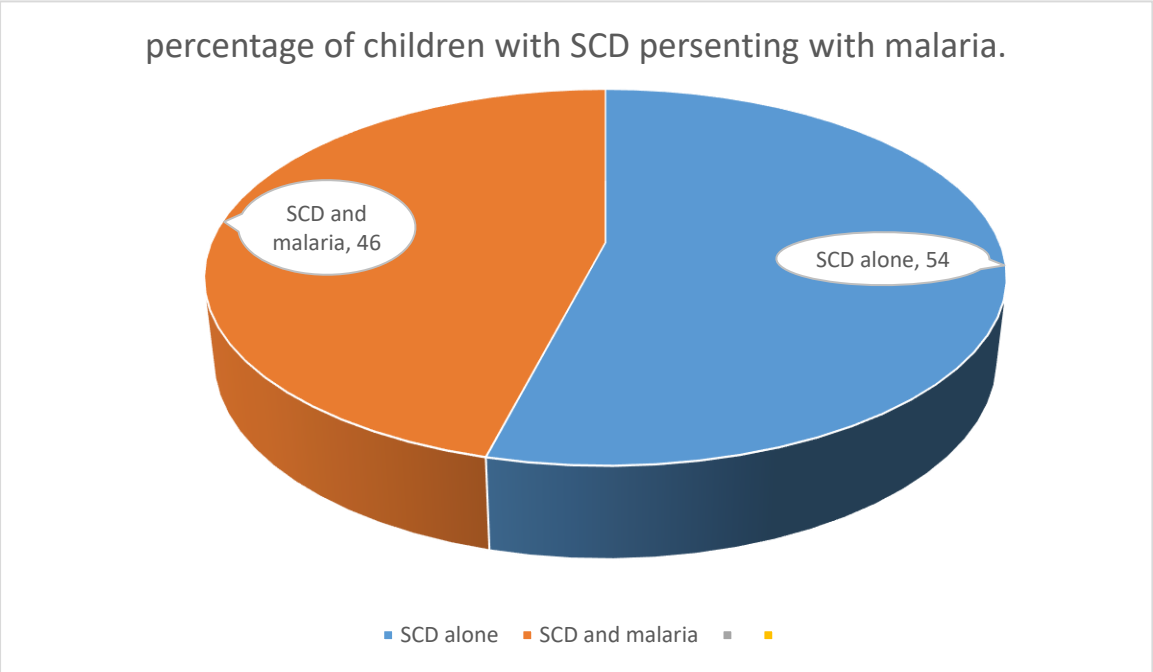
Prevalence of SCD as a percentage of the admissions bellow ten years in the unit was 2.2%

4.1.1(a) Figure 1 Pie chart indicating the Sex of the patients.



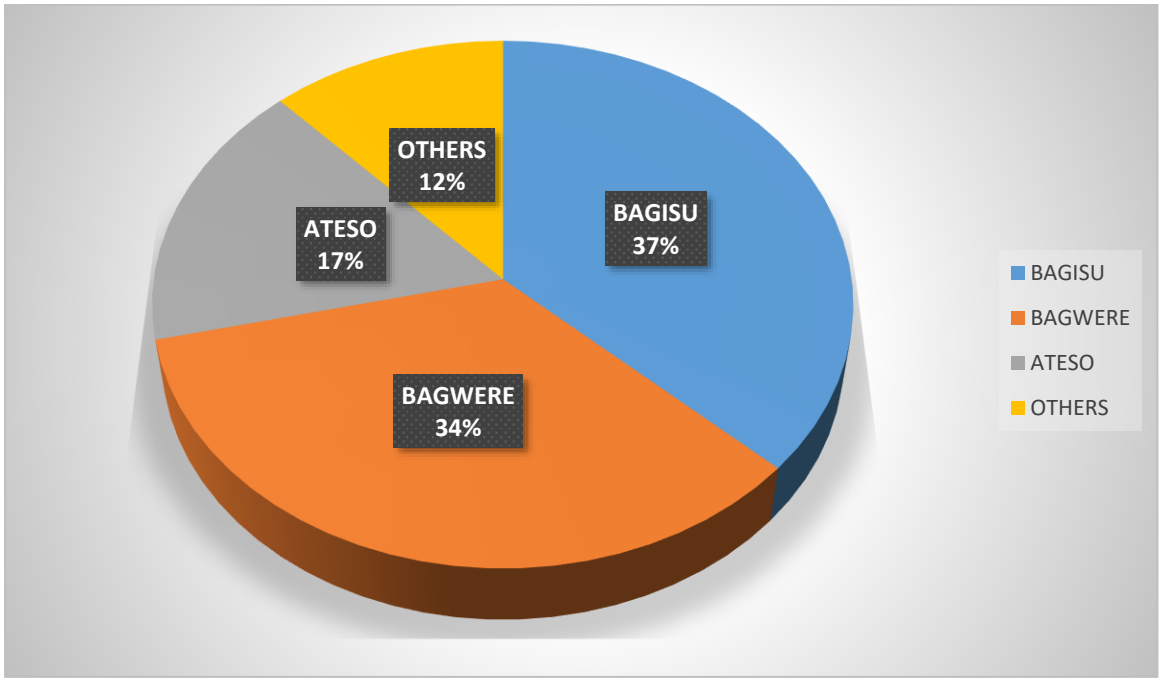
Majority of the patients diagnosed with sickle cell disease were females representing 53% while males represented 47

4.1.1(b) Figure 2 Pie chart indicating the number of patients who presented with SCD and malaria.



Majority of the admitted presented with other complaints other than malaria although it was the highest and most important complaint.

4.1.1(c) Figure 3 Pie chart indicating admissions of by ethnicity.

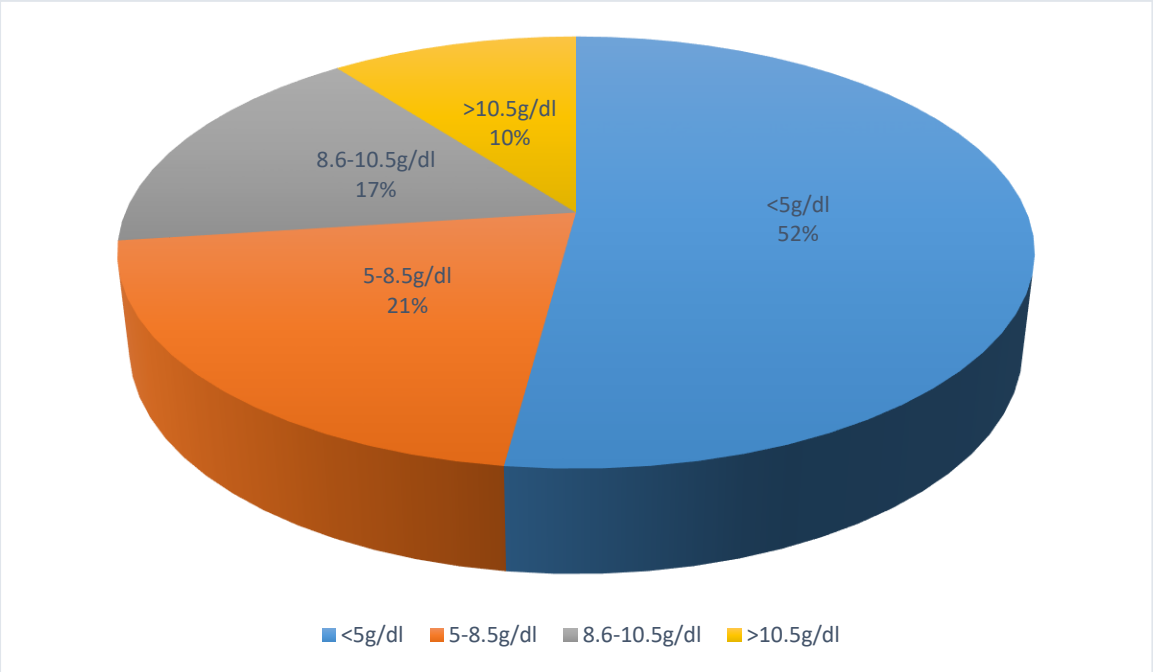


Majority of the patients admitted were predominantly Bagisu accounting for 37% followed by the Bagwere who accounted for 34%

Table 2. Summary of the observed Specific positivity among the different ethnic groups.

Ethnicity	No of admissions	Number SCD positive	Percentage positive
Bagisu	**2520**	**55.1**	**2.3%**
Bagwere	**2300**	**50.1**	**2.0%**
Ateso	**1698**	**25.3**	**1.5%**
Others	**1215**	**17.9**	**1.14%**
Total	**6790**	**149**	

4.1.1(d) Figure 3 pie chart representing haemoglobin levels with which the children presented with.



Majority of the patients admitted had low Hb 52% having between 5-8.5g/dl. Only about 10% of the children were not anaemic.

Table 3. Admission by denomination.

Denomination	No of patients	SCD positive	%of SCD positive
Catholic	2050	51	34.2%
Anglican	1860	38	25.5%
Islam	1500	31	20.8%
Pentecostal	1002	22	14.8%
Others	378	7	4.70%
Total	6790	149	100%

SCD was more common in the Catholics at 34.2% followed by the Anglicans at 25.5% its least in the others at 4.70%

Table 4. Proportion of SCD co-infection with malaria.

SCD POSITIVE	**MALARIA POSITIVE**	**PERCENT POSITIVE**
149	**70**	**47.0%**

47.0% of the patients with SCD were co-infected with malaria.

Table 5. Proportion of malaria in the general admissions.

Total no of admissions bellow 10	**Malaria positive**	**percentage**
6790	**2716**	**40.0%**

40% of the admissions bellow ten in the unit were due to malaria.

Table 6. Indicating positivity of malaria with ethnicity both general and specific prevalence.

Ethnicity	Number positive	Percentage positive
Bagisu	**1222**	**45%**
Bagwere	**841**	**31%**
Ateso	**299**	**11%**
Others	**353**	**13%**
Total	**2716**	**100%**

Specific positivity indicated that the Bagisu had more malaria positives at 45% followed by the Bagwere.

4.2 QUALITATIVE RESULTS

4.2.1 Response from the paediatric unit in-charge.

The in-charge reported that the disease on the paediatric ward in order of occurrence were noted to be malaria, pneumonia, diarrhoea, malnutrition and others including sickle cell. Among these malaria was the leading cause of admissions in the under ten years being admitted to the paediatric unit of Mbale Regional Referral Hospital. Malaria cases where by far the commonest causes of blood transfusions on the ward. Sickle cell disease also accounted for a big percentage because many of those who were admitted almost always needed a blood transfusion.

In a bid to offer preventative services, the Hospital runs a community outreach programme that does educative preventative and diagnostic services of great importance is the immunisation outreach programme that carries out vaccination of children in the villages. They also provide daily health education at the centre.

The hospital also runs a special clinic for sickle cell disease that runs on Tuesday and Thursday every week. They receive between 30 to 40 patients per week who come to receive specialised care for sickle cell disease they come from over 10 Districts that surround Mbale District. It many offers patients with prophylaxis with daily folic acid, and penicillin V to protect against pneumococcal infection. For those patients common painful crisis they also give them potent oral analgesia to go with home of which may include opiates. Most importantly they are given health education on how to prevent the different types of crisis associated with sickle cell disease.

The hospital faces a number of challenges of which include stock outs. He thinks that the stock runs out quickly because they serve beyond their catchment area and population and also that patients do not follow the normal referral systems since many do not go through the Health centres District Hospitals at the bottom of the system.

Suggestions from the unit in charge towards the prevention and control of childhood illnesses were that the government should step up its efforts to provide for the outreach services, ensure timely delivery of supplies, enhance workers allowances and then the stuff will get motivated to work.

4.2.2. Responses from the ward staff and record review.

Mbale Regional Referral Hospital admits an average of about 2500 children per month, but these numbers vary with months of the year for example numbers are very low during December.

The number of admissions contributed by malaria was is the greatest and accounts for over ¼ of all the admissions to the unit on average 600 cases of malaria are being admitted to the paediatric unit monthly some of which are readmissions.

Many of the patients admitted presented with anaemia most of which was caused by infections or infestation like malaria pneumonia. Non infection caused anaemia is mainly caused by malnutrition and sickle cell disease. Cases of children with SCD with co infection with malaria presenting with anaemia are a common finding on the ward.

The numbers of children admitted with SCD to the unit varied greatly from as many as 5 per day to none at all. The ward in charge also noted that the numbers of admissions go high during the cold or wet months of the year.

Patients are tested for SCD on suspicion based on the symptoms with which they presented with. Mothers complain of swelling of their children's fingers or toes and they are sent for testing other features with which children with sickle cell present with are irritability, symptoms of anaemia and fever in those who would have co infections with other illness.

Transfusions were very common. Most sicklers commonly needed blood based on the national guidelines. Those above 5g/dl are not transfused unless they have severe disease.

Many noted transfusions were due to malaria, others certainly due to SCD. Many times according to record review, the two also occurred together.

CHAPTER FIVE:

DISCUSSION OF STUDY FINDINGS, CONCLUSION AND RECOMMENDATIONS

5.1.1 Prevalence of sickle cell disease in the children under ten admitted.

Findings indicated that 2.2% of the children admitted had sickle cell disease (sickle cell anaemia (ss homozygotes)) this study findings present a slight deviation from the studies done previously in 2010 by Okwi et al (prevalence of 1.7%) for SS) this slight increase seen could be due to the fact that the 2010 study was not done in a hospital setting. The study shows a 0.5 increase from the 2010 study.

5.1.2 Distribution of sickle cell disease among different ethnic groups

Specific positivity among the Bagisu was 2.3% thus SCD was noted to be higher among the Bagisu than the other tribes that where admitted to the paediatric unit and this fact had previously been held according to Lenham 1949 study. Genes influence the frequency of occurrence certain diseases in different ethnic groups and this is a case in time (Davidson 2005)

Mutations do occur in all populations, but it's just that some are present in same populations with a higher frequency. This its self explains why the Bagisu are more prone than other tribes in the region. The precise reason however still remains unclear and may need more research.

The difference in the prevalence may also have been factored by the admissions on the ward where majority of the patients were Bagisu (37%). This requires that other studies be carried out among the health population to come up with specific ethic rates before we can precisely come out and say that the Bagisu are more affected than the rest. It may be possible that a study done predominantly Bagwere area would also give a higher specific positivity the Bagwere than in the Bagisu, and for this reason a different survey is recommended in the Bagwere predominated areas of the Region. Such a study would eliminate the bias of holding strictly that the Bagisu are more affected. Since the Bagisu occupy the mainland near the hospital while the others occupy the areas remote from the Hospital this affects the accessibility to the hospital thus the difference in the prevalence.

5.1.3 Levels of haemoglobin with which children of sickle cell presented.

Majority of the patients with SCD had low haemoglobin. This is in agreement with studies that had been done before (WHO 2006). The average HB for sicklers was 5.3g/dl. The calculated average of haemoglobin for the whole number admitted was 7.2g/dl.

The study considered that any Hb bellow 8.5g/dl is anaemia and in regard to this, not only the children with SCD alone are anaemic but also those without the disease. About 45.6% of the children admitted did not have anaemia as per the specifications above, while 54.4% were anaemic. This means that SCD was not the only reason for anaemia at the unit. Other diseases such as malaria and malnutrition might have been responsible.

5.1.4 Distribution of the patients by gender and age

Majority of the patients were female (54%) while males accounted for 46%. this is not in agreement with most studies that say males are more prone to the disease and death (Diallo D T 2003). The explanation for the above finding is a subject matter beyond the scope of this study.

In regard to age, the study showed that the numbers of the under 1 year were less as compared to those between other ages bellow 10 years. This finding seems to agree with the fact morbidity in children begins commonly after the child has lived the age 6 months a time at which maternal antibodies are presumed to have been depleted. Also of importance is foetal haemoglobin which is also still present in the early live this haemoglobin is said to be resistant to sickling thus no signs of SCD.

5.1.5 Sickle cell disease and malaria co-infection

Of the total patients who were confirmed positive for SCD 70 of them were co-infected with malaria representing 47.0%. This is a 1.5 times chance for one who is a sickler to get malaria and so this calls for concerted effort to prevent and to control malaria in the sicklers with the aim of reducing morbidity and mortality among SCD. Sicklers are prone to malaria and when they get the attacks they are often the severe forms of the disease. Their anaemic state predisposes them to lowered immunity and just as they are susceptible to serious bacterial infections they are also susceptible to malaria. On contrary those with the sickle cell trait are known to have resistance to malaria

5.2 Conclusion

Sickle cell disorders in children are indistinguishable both clinically and haematologically. They run in families and seen in siblings. Therefore, the children with recurrent episodes of painful attacks in abdomen, musculoskeletal pain, fever, splenomegaly, anaemia and epistaxis should be suspected of sickle cell disorders. They should be screened by simple sickling test to identify the genetic disorder. Parental counselling and preventive measures like penicillin prophylaxis, regular folate supplementation, early treatment of acute respiratory infections with simple antibiotics and management of pain with simple analgesics will be helpful in decreasing morbidity and mortality in childhood with sickle cell disease, hence With a comprehensive medical care and management approach, the health status and life expectancy of these patients can be improved considerably.

The study finding indicated that the prevalence of sickle cell disease (haemoglobin ss) among the under 10 admitted between May 2014 and July at Mbale Regional Referral Hospital was 2.2%

SCD is found higher in the Bagisu than the Bagwere with specific ethnic positivity rates of 2.3 and 2.1 respectively.

Aneamia was pronounced among children presenting with sickle cell disease. The average haemoglobin for the sicklers being 5.2g/dl.

Malaria ranked the number one cause of admission on the ward for both those with SCD and those with out at 40% and 47% respectively.

5.3 Recommendations:

5.3.1 National recommendations:

As Mbale is one of the districts noted to be among the areas with high SCD, government should consider offering a better method for diagnosis for example an electrophoresis machine to capture both the carrier and the sicklers.

The ministry should consider a constant supply of mosquito nets to SCD patients as this will reduce on the morbidity and mortality due to sickle cell disease.

The SCD association of Uganda in collaboration with the ministry of health Uganda should spear head the expansion of the SCD clinic in Mbale Regional Referral Hospital. This will considerably reduce the number of children who succumb to the disease at an early age.

SCD is often worsened by infections notably pneumococcal pneumonia, government should introduce the pneumococcal vaccine in Mbale.

5.3.2 Recommendation to the district:

A survey needs to be carried out in all the neighbouring districts. The next survey to be done should concentrate in areas predominantly inhibited by the Bagwere as well, to find how this would compare with previous study findings of the prevalence of the disease among the Bagisu. This shall reduce the bias of whether the results reflect the ethnic proportions.

5.3.3 Community and house hold recommendations:

Members of families affected by SCD should be encouraged to have health seeking behaviour, as their loved ones are very delicate and need proper attention when they fall sick and also because they fall sick often. Malaria was seen to be great importance in causing morbidity in these children it's there for advised that they all sleep in insecticide treated mosquito nets.

Intermarriages should be encourage as it's a good intervention in places of the country like this one where the prevalence of sickle cell disease is very high. Couples from both families with a positive history of sickle cell disease should seek marriage counselling before marriage and if possible they should go and carry out tests to determine the risk of having a sickle cell affected child.

The community should also be taught on the dangers of stigma against these children especially at school.

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Appendix I: Consent Form

FACULTY OF CLINICAL MEDICINE AND DENTISTRY,
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P.O.BOX71, BUSHENYI, UGANDA.
Tel:+256485443629
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Topic: DETERMINING THE PREVALENCE OF SICKLE CELL DISEASE AMONG CHILDREN UNDER 10 YEARS ADMITTED AT MBALE REGIONAL REFERRAL HOSPITAL BETWEEN MAY 2014 AND JULY 2014.

Investigator: WAMBI IVAN

Dear Participant

I am a research investigator from Kampala international university western campus Bushenyi district. Am conducting a study to determine the prevalence of sickle cell disease in this district. Am requesting you kindly to spare me same time and I ask you a few questions related to the topic above. The information you give to me will be treated as confidential. I am not going to take your names for record purposes.

You are free to join the study or not to join, you may leave the study at any point for any reason if you decide not to join or drop out, you will not lose any healthcare service you are entitled to at the hospital, you will not get any direct benefit or payment for being in the study, however you will help us know more about the disease condition.

The study has been explained to me and I have chances to ask questions. I've been informed that it is my free choice to be in the study

If you agree to participate in the study please sign here…… Thank you very much

Appendix II QUESTIONNAIRE TO WARD STAFF

1. How many children do you admit monthly?

   ---

2. Of these how many are due to malaria?

   ---

3. How do you conform the presence malaria?

   ---

4. How many of these present with anaemia?

   ---

5. How many present with sickle cell disease?

   ---

6. How do you conform the presence of SCD?

   ---

7. What is the commonest presentation of SCD on this ward?

   ---

8. How many are transfused?

   ---

9. Of the transfused how many are due to malaria?

   ---

10. How many are due to SCD?

Appendix III

QUESTIONNAIRE SCHEDULE TO THE HEALTH UNIT INCHARGE

1. What are the most common diseases among children under five in this area?
2. Which disease is the leading cause of admission in this facility in the less than 10 years admitted to the facility?
3. Which measures are being done to combat the major health conditions in the area?
4. What difficulties are being by the community in accessing health facilities in the area?
5. Which disease condition is the leading cause of blood transfusion among the under ten in this facility?
6. What suggestions would you put forward for the control and prevention of child hood disease in the area?

Appendix IV: Research Budget

S/No.	Items	Quantity	Rates Each	Total Amount(UG.SHS)
	Stationary			
1	Pens	10	600=	6,000=
2	Reams of paper		10000=	10,000=
3	Box files	2	5000=	10,000=
4	Note books	4	3,000=	30,000=
	Secretarial services			
1	Typing, printing and binding of research report	5 copies	20000=	100,000=
	Others			
1	Food/water	4 days	10,000=	40,000=
2	Transport	2days	40000	80,000=
	TOTAL			**276,000=**

MAP OF UGANDA SHOWING MBALE DISTRICT

POSITION OF MBALE