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Assessing Knowledge Level, Perception, and practices among policy-makers toward Sickle Cell Disease in Kalangala, Uganda

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Assessing Knowledge Level, Perception, and practices among policy-makers toward Sickle Cell Disease in Kalangala, Uganda

Directed by Professor Min Jin Ha

A Master's Thesis

Submitted to the Department of Global Health and Disease Control
Division of Health Policy and Financing
and the Graduate School of Public Health of Yonsei University
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Anthony Byarugaba

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DECLARATION

I dedicate this work to the memory of my parents, Monica Ateenyi Kyomuhendo and the late Dr. Baraba Patrick Akiiki, who has long since departed to a higher realm. Your kindness, decency, diligence, and honesty taught me as a child all I ever needed to know about what a good man should be. I'm grateful for that strong base.

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I value the knowledge Yonsei University helped me acquire while pursuing a master's degree. Yonsei University's ideals have profoundly changed my life, and I will always be grateful for their assistance in realizing my dream!

My greatest gratitude is to my Alpha and Omega, who have never turned away from me or withheld His love."Amen, glory be to His holy name."

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LIST OF ABBREVIATIONS

SCD	Sickle Cell Disease
WHO	World Health Organization
MOH	Ministry of Health
DHO	District Health Officer
H/W	Health Worker
NGO	Non-Government Organization
N/A	Not Applicable
CSO	Civil Society Organization
UN	United Nations
CDC	Center for Diseases Control
SCA	Sickle Cell Anemia
DHIS2	District Health Information System
RBC	Red Blood Cells
LMICs	Low Middle-Income Countries
DBS	Dried blood spot
CPHL	Central Public Health Laboratory
POC	Point of Care
HBM	Health Belief Model
SCT	Social Cognitive Theory
CI	Confidence Interval

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ABSTRACT

Globally, Sickle cell disease (SCD) is a congenital blood illness that affects millions of people. Uganda has the 5th highest burden in Africa, (Thomson et al., 2023). The health policies that affect the diagnosis and management of SCD are heavily influenced by policymakers. Good knowledge and understanding of SCD amongst Policymakers substantially impact support and resources allotted to fighting the disease with reduction of the prevalence.

The study Assesses Knowledge, Perception, and practices among policymakers toward SCD in Kalangala Uganda.

A cross-sectional survey was undertaken among policy-makers residing in Kalangala. 194 Google forms sent to participants using convenience sampling and snowball method. A total of 151 Google forms returned with a response rate of 77.83%. The analysis was performed using the Jamovi-2.3.21.0 version. Descriptive, Chi-square tests, statistical analyses were carried out. Female respondents were in the majority as shown by 58.9% of the sample, and the dominant age group was 25-34 years (51.0%). Most of the respondents (95.4%), ever heard of SCD. Some misconceptions were identified, such as, believing that praying could cure sickle cell disease while others believed someone with SCD couldn't work.

This study identifies knowledge gaps about SCD which results in poor health outcomes (Martinez et al., 2020). In a society with a high sickle cell population, there is a need for more common sense regarding how to prevent and manage the risks of SCD. Additional research supports the use of sickle cell disease (SCD) in health education campaigns as part of contextual strategies by policymakers, stakeholders, and government to prevent SCD.

Keywords: Sickle cell disease, Knowledge, Practices, perceptions, policy-makers

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I. INTRODUCTION

1.1 Background

The Uganda Sickle Surveillance investigation carried out a cross-sectional investigation that revealed a high frequency of sickle cell trait and disease in Uganda, with substantial regional and district-level variations. In Uganda, the illness poses a concern to public health. About 25% of Ugandans are carriers, which represents 13.3% of the country's population (Tusuubira et al., 2018). SCD is a chronic infection, characterized by recurrent organ damage, and shortened life span. It’s necessary for Sickle Cell Disease control through public health education and other preventive measures. Sickle cell disease was a widely recognized condition in West Africa, where it was known by many local names. (Reid & Rodgers, 2007). Millions of people worldwide suffer from SCD, which is the most common blood condition in families with ancestors from the Mediterranean, South America, Caribbean, Saudi Arabia, and Africa. (Creary et al., 2007).

1.2. Problem statement

Globally, the burden of sickle cell disease (SCD) has not been sufficiently addressed. As a result of improving longevity in high-prevalence low- and middle-income countries, the burden of sickle cell Disease (SCD) is expected to increase globally. Twenty million people worldwide and more than 100,000 Americans are affected by the illness (Carden & Little, 2019). In sub-Saharan Africa, sickle cell disease is a significant public health concern acknowledged as a major cause of morbidity and infant mortality. It is a significant hereditary disease in the majority of Sub-Saharan African nations. However, in several nations, like Cameroon, the Democratic Republic of Congo, Gabon, Ghana, and Nigeria, the prevalence is between 20% and 30%. Overall in Uganda, Sickle cell trait prevalence was 13.3%, but in eight districts, it exceeded 20%. More than 15000 babies

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are born with the disease every year, according to data on the total prevalence of sickle cell trait (13.2%) and disease (0.8%) among infants 6 months of age or younger. Some districts of Uganda, stand at 45% (Ndeezi et al., 2016). In Africa, sickle cell disease dramatically raises the mortality rate for children under five, which makes it more difficult to accomplish UN Sustainable Development Goal No. 3: Good Health and Well-Being, which calls for a reduction in childhood mortality.. According to the CDC Sickle Cell report, Uganda is Africa's fifth-highest burden, which is made worse by the lack of established social support systems for clients plus their family members to help them deal with the emotional effects of SCD. When the disease first manifests, the affected person's life expectancy and quality of life are drastically decreased (Machnik Sickle Cell Foundation initiative; Global Fund for Forgotten People, December 2018). Compounded with the stigmatization of people with sickle cell illness, isolating them away from their family and community members making them vulnerable owing to Uganda's high malaria load (over 18%). In a significant report on sickle cell disease in Africa, the World Health Organization (WHO) outlined the disease's overall prevalence and offered recommendations for raising awareness of the condition and enhancing its prevention, early detection, treatment, and management approaches. Among the major challenges facing health ministries is the dearth of precise information regarding the prevalence and geographic distribution of the illness in their nations (Diop & Pirenne, 2021).

Furthermore, in Uganda, SCA is thought to be responsible for 16.2% of all pediatric fatalities. In Uganda, it's estimated that around 20,000 infants are born with sickle cell disease each year, while precise figures are lacking (Ndeezi et al., 2016). SCD clinic in Kalangala has a total of 74 patients on care and management (DHIS2 2022) and Kalangala ranks first in Uganda for highest HIV and malaria prevalence at 18.8% and 18.2% respectively compared to the national of 5.1% and 19% (CDC Nov 2021) puts it to the highest prone of the SCD. While SCD-targeting therapies and technology are becoming more widely accessible in Uganda, many of the country's leaders and policy

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officials are unaware of the disease's symptoms, potential treatments, acceptability, and practicality of axillary therapy.

1.3. Purpose

This was to evaluate Ugandan policymakers' knowledge, perceptions, and practices regarding SCD. Sickle cell disease (SCD) is the most confusing infection in the community and one of the blood disorders and chronic diseases with the least understanding.. Additionally, current SCD research focuses on medical students' knowledge of the illness in institutions and Universities, causing a lack of information regarding the awareness among policy-makers, and the community. Furthermore, I tried to assess how policymakers' knowledge would be used for SCD Policy reviews.

1.4. Objectives

- To assess the awareness and understanding of Sickle Cell Disease among policy-makers in Kalangala district.
- To associate knowledge level, perception, and practices among age groups, gender, education level, marital status, and having biological children regarding sickle cell disease.
- To propose recommendations and strategies to improve the knowledge, perception, and practices of policy-makers in Kalangala, Uganda regarding Sickle Cell Disease based on the findings of the study.

1.5. Hypotheses

There is an association between the level of knowledge regarding Sickle Cell Disease among policy-makers based on their age groups, gender, marital status, educational levels, and having biological children.

The perception and practices of policy-makers towards Sickle Cell Disease are

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influenced by their age groups, gender, marital status, educational levels, and biological children.

1.6. Definitions

Sickle cell disease (SCD): Is a genetic blood disorder caused by abnormal hemoglobin that damages and deforms red blood cells (RBCs) (Creary et al., 2007).

Sickle cell trait: one defective gene and one healthy gene inherited together. Although sicklers typically don't exhibit any symptoms, they can nonetheless pass the characteristic on to their progeny (CDC, 2017).

Policy; Government initiatives and programs, both current and planned, aimed at addressing many societal issues. Public policy, then, is the result of authoritative or approved decisions made by government actors. (Nadel, 1975).

Genetic counseling is a method of communication that addresses the issues that people face when a genetic illness runs in the family or has the potential to run in the family. (Rantanen et al., 2008).

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II. LITERATURE REVIEW

2.1. Introduction

This summarizes the published research on policymakers' roles in health promotion in the literature review. The major objective of the study was accomplished by retrieving journal articles from multiple databases, such as PubMed, MEDLINE, and Google Scholar.

2.2. Sickle Cell Disease (SCD)

In 2022, hematologists' contacts with Medicaid recipients characterized sickle cell disease as a hereditary blood disorder that affects hemoglobin, the red blood cell protein responsible for carrying oxygen throughout the body (Horiuchi et al., 2022). Even if the life expectancy of people with sickle cell disease (SCD) has increased recently, the disease's cumulative consequences cause serious sickness and lower quality of life for many. Acute and chronic pain episodes, the risk of organ damage and stroke, and a high acute health care consumption rate are common symptoms of sickle cell disease (SCD). A thorough summary of sickle cell disease can be found in the review of the literature. This review covers the history of sickle cell disease, its complications, treatments, campaigns to raise awareness of the condition, genetic testing, newborn screening, the patient-physician relationship, and public perceptions of the condition among policymakers, healthcare professionals, and the general public. This review's main objective is to draw attention to the fact that sickle cell disease policymakers are largely unaware of the condition. This literature review examines the current state of knowledge, perception, and practices surrounding SCD among policymakers. Studies have consistently shown that policymakers often have limited knowledge and understanding of SCD. A study conducted in Nigeria found that only 16% of policymakers had a good knowledge of

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SCD, while 67% had poor knowledge (Olatona et al., 2012). Similarly, a study in the United States found that only 16% of state health department directors felt confident in their knowledge of SCD (Phillips et al., 2022). This lack of knowledge can lead to a lack of prioritization and underfunding of initiatives to address the disease. Perceptions and attitudes towards SCD among policymakers can also contribute to stigma and discrimination, which can further hinder effective disease management. A study conducted in Ghana found that policymakers often held negative attitudes toward individuals with SCD, including beliefs that the disease was caused by witchcraft or immoral behavior (Dennis-Antwi et al., 2019). Such perceptions can lead to inadequate policies and programs, delayed diagnosis, and poor health outcomes for individuals living with SCD. Efforts to improve knowledge, perception, and practices surrounding SCD among policymakers have included advocacy initiatives, increased awareness campaigns, and education and training programs. In the United States, advocacy organizations have worked to raise awareness of SCD and its impact on individuals and families. A study conducted in the capital of Kampala indicates that, Out of 110 individuals surveyed, 91.2% of them had heard of sickle cell disease (SCD), with the highest proportion (38.7%) learning about it from friends and family. Almost half of the respondents (48%) knew that SCD is an inherited condition, but a significant proportion (44.2%) were unaware of its cause. However, 68.7% of the respondents stated that they would not marry someone with SCD. According to (Tusuubira et al., 2018), raising awareness about SCD is an important strategy for preventing the disease as it helps people to make informed choice about the marriage. Therefore, evaluating policymakers' sickle cell disease knowledge can aid in the development of pertinent public health policies and initiatives that raise awareness and knowledge of the illness. Thirty percent of the African Americans these authors contacted were dropped from the study because they had no prior knowledge of SCD. Out of 162 respondents who satisfied the requirements, 9.3% were aware of the disease pattern. 11% had no idea if they were carriers (Boyd et al., 2005). Participants also lacked knowledge about SCD treatment approaches, even though

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the majority were aware that pain episodes are a significant complication and that SCD is a faulty blood disease. (Boyd et al., 2005). Strong evidence was found in this study to suggest that African-American women in their prime reproductive years still lack knowledge about the signs, prevalence, and inheritance patterns of sickle cell disease. (Boyd et al., 2005) in SCD. Making decisions about having children requires having a general awareness of sickle cell disease. In order to determine the knowledge and exposure of 282 people from Northern California to sickle cell disease and sickle cell characteristics, Treadwell et al. (2006) conducted a survey (Treadwell et al., 2006). It's interesting to note that 68% of study participants who were interviewed gave accurate answers to SCD knowledge questions. Merely 15% of the participants acknowledged their characteristic status. Thirty respondents, in the main, stated that they had SCD testing before. Ogamdi (1994) assessed 334 students from a southeast Texas university's general SCD knowledge. 81% of students knew the gene that causes sickle cell disease (SCD), however more than 60% were unaware that SCD is a condition that may be prevented if people make "responsible" reproductive decisions, according to study data (Ogamdi & Onwe, 2000). On the other hand, 63% of pupils correctly answered knowledge questions regarding the symptoms of SCD (Ogamdi & Onwe, 2000). The fact that not all questionnaire answers were provided in frequency was a major study drawback. However, this researcher concluded that people in the 19–30 age range don't know the fundamentals of sickle cell disease. There is a need for more sickle cell tests, education, and counseling among university students notwithstanding the existing rift among policymakers.

A similar survey, 191 black American students from the Southeast, aged 19-30, were the sample for a mixed-method study carried out by Stewart and colleagues (Stewart, 2007). Their research examined young black American adults' knowledge and beliefs about sickle cell disease (SCD), the SCD trait, and genetic testing (Stewart, 2007). The target group lacked an understanding of genetic testing, family history, and carrier status, the investigators discovered (Osbourne, 2012). Lastly, the knowledge and awareness of

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SCD were assessed for 104 SCD carriers and non-carriers in a research study conducted by (Boadu, 2018).

2.3. Genotypes and Pathophysiology of SCD

A single-point mutation in the HBB gene results in a set of crippling autosomal recessive illnesses known as sickle cell disease (SCD). The mutation at position 6 on the hemoglobin beta-globin subunit, which is encoded by the HBB gene, converts a hydrophilic glutamic acid to a hydrophobic valine. Other names for this mutation include p.Glu6Va and c.20A>T. Sickle hemoglobin (HbS), an abnormal form of hemoglobin, develops as a result of this mutation. Traditionally, SCD is inherited in an autosomal recessive Mendelian pattern, as seen in Figure 1 below, where a male or female kid with the disorder inherits one defective gene from each parent;

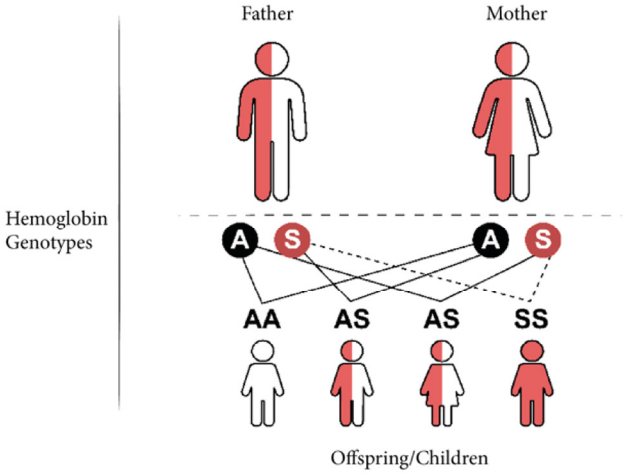

Figure 1. Showing Inheritance of sickle cell disease
(**Source:** Cooley, T. B., & Lee, P. (1926). The sickle cell phenomenon. American Journal of Diseases of Children, 32(3), 334-340.)

If both parents have sickle cell trait (SCT, HbAS), there is a 25% probability of having a normal offspring (HbAA), a 50% risk of creating an offspring with sickle cell trait

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(SCT), and a 25% chance of producing an offspring with sickle cell disease (HbSS) with every pregnancy. Attributed to the inheritance of two copies of the HbS mutation (homozygotes), sickle cell anemia (HbSS) accounts for 65–70% of all instances of sickle cell disease (SCD) and is the most common genotype. The additional common types of sickle cell disease (SCD) that arise from the coinheritance of mutations that cause other abnormal types of hemoglobin (like HbC or -thalassemia) in combination with HbS, forming a compound heterozygous mutation, are hemoglobin SC (HbSC), sickle hemoglobin-beta-zero thalassemia (HbS0thalassemia), and sickle hemoglobin-plus thalassemia (HbS+thalassemia). Compared to individuals with HbS0thalassemia, who generate no -globin at all, those with HbS+thalassemia produce less -globin. The identical site (c.19G>A, p.Glu6Lys) where glutamic acid is substituted with lysine in HbS is the cause of HbC. Other -globin gene variations include HbE, Hb-Lepore, HbO-Arab (c.364G>A, p.Glu121Lys), Hb Quebec-CHORI, and HbD Punjab/Los Angeles (c.364G>C, p.Glu121Gln). The coherence of HbS and these hemoglobin variants result in rare and often benign SCD genotypes (HbSD-Punjab/Los Angeles, HbSE, HbS-Lepore, HbSO-Arab, and HbS-Quebec-CHORI).

2.4. Impact and Complications of Sickle Cell Disease

It has been demonstrated that providing people with SCD access to comprehensive, high-quality care improves outcomes and decreases acute care utilization (hospitalizations and ER visits), especially among adults. Although centers in some regions of Uganda offer complete care for children with SCD, such centers are uncommon. Adult healthcare practitioners who specialize in non-malignant hematology are also uncommon. Adults who leave pediatric care run into a shortage of specialists who understand SCD and a disjointed healthcare system. Even the most straightforward of these models that of a patient being managed by a hematologist and primary care physician, is infrequently observed in practice. Several models have been proposed for best practice management for SCD.

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A study carried out in Uganda found that sickle cell trait and disease were highly prevalent, with noteworthy regional and district-specific variations. According to the study, 0.7% of people had sickle cell illness and 13.3% of people had sickle cell trait. The study also found that sickle cell disease contributes substantially to mortality in children younger than 5 years in sub-Saharan Africa, and in Uganda, 20,000 babies per year are thought to be born with sickle cell disease. The percentage of the SCD population that does not receive care is unknown due to a lack of data and limited access to health services.

Stroke one of the many tragic side effects of SCD that can result in death or lifelong brain damage in children is a stroke. A major disturbance in the delivery of oxygen to the brain causes a stroke, which results in a reduction in oxygen flow. This difficulty could develop suddenly. According to studies, a stroke in an SCD patient typically occurs when they are 6 years old. Studies have also shown that at least 8% of SCD patients have experienced a stroke by the age of 14, and 11% of SCD patients have experienced a stroke by the age of 20 (Osbourne, 2012). For 3years of SCD treatment, children had 50-70 percent chance of experiencing another stroke (Thomas et al., 1997). Stroke diagnoses can be made using magnetic resonance imaging (MRI) and transcranial Doppler ultrasonography (TCD). The body of research suggests that there is a need for a stroke diagnostic approach that is more precise than TCD. Furthermore, studies show a lack of educational materials about the consequences of strokes, and these findings point to the necessity of expanding stroke screening programs. The magnitude of this problem was shown by a cross-sectional study conducted by The Children's Hospital of Philadelphia (CHOP) with 44 participants. The study found that 46% of caregivers surveyed were unable to identify stroke warning signs. Merely 34% of the caregivers in this study were able to identify a stroke as a consequence of SCD (Katz et al., 2002). Acute chest syndrome (ACS) is a severe side effect of sickle cell disease (SCD) that increases rates of morbidity and death. The ACS is brought on by lung cavity bacterial infections. Reports state that over 50% of individuals with sickle cell disease (SCD)

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experience ACS, a potentially fatal result similar to pneumonia. Recurrence of ACS has been noted in approximately 80% of SCD patients (NAMAZZI et al., 2022). About 25% of untimely mortality among SCD patients is attributed to Acute Chest Syndrome (ACS). No current laboratory, radiographic, or clinical techniques can reliably determine the cause of ACS. The most obvious causes of ACS, according to researchers (Yousef et al., 2022), include infections and fat emboli. Fever, coughing, abnormal chest X-rays, and chest pains are the most typical diagnostic signs across all age groups. Wheezing, a productive cough, and shortness of breath are less frequent symptoms (Yousef et al., 2022). In 1998, the FDA granted a license for the use of Hydroxyurea, a well-known cytostatic myelosuppressive chemotherapy drug, to adult sickle cell disease patients. (Kambasu et al., 2019). Hydroxyurea was a chemotherapeutic medication used for many years in the treatment of cancer. These days, it's utilized to decrease erythrocyte sickling and sickle cell hemoglobin polymerization and boost the formation of fetal hemoglobin (HbF), a hemoglobin found in infants (Kambasu et al., 2019). Hydroxyurea therapy has been effective in reducing the frequency of painful episodes and the recurrence of common sickle cell disease (SCD) complications. Patients who experience painful episodes on a regular basis or who have a history of acute chest syndrome, symptomatic anemia, or other vaso-occlusive events are advised to take hydroxyurea medicine by medical specialists (Nnebe-Agumadu et al., 2021). Hydroxyurea treatment significantly decreased the incidence of acute chest syndrome and vaso-occlusive crises in 19 adults with hemoglobin SS enrolled in a random placebo-controlled trial research (Nnebe-Agumadu et al., 2021). The study found that the number of unpleasant events fell from 4.5 to 2.5 per year. After 28 months of treatment, hydroxyurea's double-blind trial, which had 299 adult sickle cell disease (SCD) patients in placebo-controlled trials, was terminated due to the drug's ability to lower patients' incidence of acute chest syndrome events and pain episodes. (Steinberg, 1999). The National Heart, Lung, and Blood Institute (NHLBI) conducted a pilot research to assess hydroxyurea's potential benefits for pediatric patients. Younger children have good tolerance to the medication, as

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evidenced by the participants, who ranged from 6-24 months. According to scientists, this medication will play a significant role in developing nations. Many impoverished nations lack the infrastructure required to carry out bone marrow infusions and transplants in a safe manner.

Medicine by Transfusion is an additional tactic for managing SCD issues and helping to prevent strokes and acute chest syndrome. According to (Natukunda et al., 2010), Patients with sickle cell disease (SCD) who have heart failure, severe anemia in children, enlarged spleens, aplastic crises, splenic sequestration, multi-organ failure syndrome, or who are stroke-prone are advised to have transfusions. Increased hemoglobin A and decreased sickle hemoglobin percentage are the goals of transfusion therapy, which aims to improve blood's ability to carry oxygen. (Natukunda et al., 2010) According to a clinical trial study assessing the efficacy of transfusions in patients with specific SCD aftereffects, children with SCD who get repeated transfusions are less likely to experience subsequent strokes. It also demonstrates that, in comparison to SCD patients who had received transfusions, only 10% of children with strokes who had not had transfusions were predicted to experience strokes within three years. Bone Marrow Transplantation Bone marrow transplantation is an experimental therapy limited to candidates below 16 years of age who exhibit SCD complications, such as a stroke, acute chest syndrome, and refractory pain. Statistics show that only 1% of patients with sickle cell anemia meet the set requirements for bone marrow transplantation (Howard et al., 2015).

A transfusion of bone marrow was utilized in 1984 to manage a leukemia client in a pediatric group. The patient's sickle cell illness was also cured by the transplant (NIH, 2002). Researchers have discovered that 10% of patients die from this treatment and that patients with sickle cell disease must first complete a chemotherapeutic program before receiving a transplant. As a result, some patients die during chemotherapy phase, while others develop fatal infections before adequate regeneration of their immune systems and bone marrow (NIH, 2002). Graft versus host disease (GVHD) and graft rejection are possible side effects. At the moment. Researchers at the National Heart, Lung, and Blood

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Institute (2002) are investigating ways to lower the danger associated with bone marrow transplantation. It is thought that a baby's umbilical cord blood lowers the likelihood of graft-versus-host disease and rejection. Furthermore, unlike bone marrow donors, this blood does not have to closely match the recipient's blood type (NHLBI, 2002). Experimental Medical Care a novel treatment for sickle cell anemia clients is gene therapy. Because gene therapy provides a more long-lasting answer than blood transfusions, researchers feel that it is preferable to the latter. A delivery mechanism that can transfer a healthy gene to cells with damaged genes is necessary for gene therapy (Leibovitch et al., 2022). Modified viruses have been selected by scientists as an effective delivery mechanism. To make sure that the normal genes work as they should, normal genes are sent to the target cells. Although research on gene therapy is still in its infancy, many scientists think that this approach may eventually lead to a clinical cure for sickle cell anemia.

2.5. Factors Associated with Sickle Cell Disease

2.5.1. Genetic Cause of Sickle Cell Disease

According to Serjeant (2013), sickle cell disease (SCD) is caused by inheritance of the sickle cell gene. Individual hemoglobin genes cause the SCD. Homozygous SCD is the most prevalent genotype that is present at birth. This particular genotype frequently shows up as a higher death rate. The geographic distribution and age of each gene have an impact on the genotype as well (Muganyizi, 2013 #78). Serjeant (2013) asserts that SCD is Central Africa, Benin, and Senegal. Ellithy, Yousri, and Shahin (2015) also emphasized that genetic and environmental variables together account for the majority of the variance in SCD. Ellithy et al. have made note of the fact that physiological changes, such as several genetic mutations and polymorphisms, might influence how SCD manifests. Glutathione S-transferase (GST) hormones with inherited polymorphisms can have insufficient enzyme activity (Ellithy et al., 2015). The GST gene polymorphism may

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cause GSH to lose its protective function against potential oxidative stress, which may in turn cause the appearance of SCD (Ndeezi, 2016 #37). SCD is referred to as a genetic condition because it is hereditary, according to Nelson (2011). If a child inherits two faulty genes (i.e., one from each parent), they are born with SCD. The morphology of red blood cells can change as a function of inherited genes (Nelson, 2011). The disease affects hemoglobin, a protein present in red blood cells. People with sickle cell disease (SCD) have hemoglobin S (HBS; Nelson, 2011). Red blood cells become sticky and brittle as a result of the HBS molecules clumping together (Nelson, 2011).

2.5.2. Current State of Newborn screening for SCD in Uganda and other countries

To lessen the burden of disease in Uganda and other countries in the region, programs for newborn sickle cell screening and improved SCD treatment capabilities are being developed. A group of key stakeholders were interviewed to gain insights into public health and international leadership perspectives, as well as the perspectives of patients and families. The interviews also sought to comprehend national progress, resource requirements, social and medical hurdles to program efficacy, and HIV/AIDS-related resources.

Collaboration between program leadership, specialists, patients and their families, international stakeholders, and the use of program resources are all necessary for successful programs. Following screening, the healthcare system must contend with a number of interconnected issues, such as delivering care by qualified medical professionals, informing parents, connecting impacted children to prompt medical follow-up, removing social and financial barriers to parental acceptance, providing education and counseling, and gaining community acceptance of the advantages of screening and care. Gaining an understanding of these elements is essential to providing impacted children with the resources they require.

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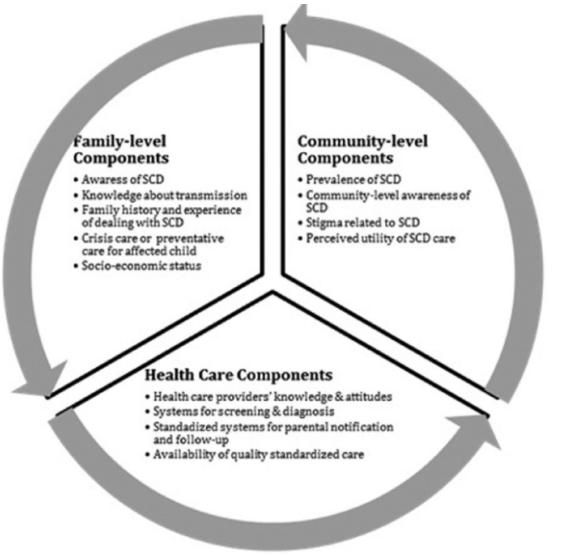

Figure 2. Shows key components of public health response to SCD

The initial stage in the active therapy of SCD is the early detection of this medical condition through neonatal screening. The UN identified sickle cell anemia as a public health issue in 2008. WHO established SCD plan for WHO African Region in 2010, including neonatal screening. Some low- and middle-income countries (LMICs) have initiated sickle cell disease (SCD) screening programs, including Uganda, where the prevalence of SCD was reported to be 0.73%, and the prevalence of sickle cell trait was reported to be 13.3% in 2015. To address this, Uganda incorporated newborn SCD screening into an existing program that identifies and provides care for HIV-positive mothers and their exposed infants in high-HIV load regions. As a component of this SCD program, dried blood spot (DBS) samples are taken from newborns at medical facilities and prior to their release, as well as from children younger than two years old at medical facilities during immunization appointments, inpatient stays, or outpatient stays. The Central Public Health Laboratory (CPHL) receives samples for isoelectric focusing (IEF) examination. The early infant diagnosis (EID) program's sample transport network is utilized to transfer DBS samples for the SCD newborn screening program. This screening

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method needs a reliable logistical infrastructure to deliver samples to and from the CPHL, as well as a functioning and financially supported system to inform carers of the results. The logistical challenge of delivering samples to and results from the CPHL can have a detrimental effect on remote facilities. Early detection, education, and management of sickle cell disease (SCD) in infants and children would be made possible by performing point-of-care (POC) testing for newborn screening at primary health facilities and giving results to caregivers during the same visit. This would more successfully prevent early mortality. There are currently initiatives trying to close this technological gap by creating POC tests for low-resource environments. There are already a few SCD POC tests available for purchase. Sickle CAN®, a multiplexed qualitative POC immunoassay utilized for the quick diagnosis of SCD, is now available commercially from BioMedomics, Inc. in Durham, North Carolina, USA, for about $4.50 per test. Laboratory testing on 139 samples of venous blood utilizing this test has shown good findings, including high sensitivity and specificity for the identification of HbA, HbS, and HbC. A disposable test chip and a small reader called Heme-Chip (Hemex Health, Portland, OR, USA) are used to analyze blood using cellulose acetate electrophoresis. These Point-of-care (POC) tests are, however, relatively unavailable in LMICs like Uganda. It is still unclear what the target cost of an SCD POC test should be for public sector programs in LMIC to be able to afford it. Target product profiles (TPPs), which technology developers should create in the early stages of their product development process to ensure alignment between Product performance and operational characteristics with end user's needs, include the target price as a characteristic. Unfortunately, the necessary rigor is not often applied to this crucial part of early product development. The price for a product used has frequently been determined through qualitative assessments, such as expert interviews and discussions. Additionally, past purchases of comparable products and currently available competitor products by potential customers are occasionally taken into consideration. However, it's crucial to take into account both the market pricing and the economic costs of implementing the new technology in a

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programmatic context when estimating the cost for an item. These economic costs comprise both direct expenses, and indirect expenses. There are few cost evaluations of newborn SCD screening in LMIC. According to one study, the cost per infant examined for a newborn screening program in Angola that included DBS was $15.36 in international currency in 2011.

Another study projected that adopting DBS for screening would cost $9.94 in Uganda, however, it's not clear how these prices were calculated {Mvundura, 2019 #83}. None of these studies evaluated the financial expenses of DBSs processed using IEF versus POC testing. We thus conducted costing research to determine the financial and economic cost per child tested and diagnosed for SCD under an existing program in Uganda. This study aims to provide better guidance for establishing the target price of the SCD point of care. Actual outlays for products and services are represented by financial costs, which are typically taken into account throughout the budgeting process. Opportunity costs are the price paid for not using already available resources, such as government employees' human resource time. The total of the financial and opportunity expenses is the economic cost.

2.5.3. Health System Capacity of SCD Clinical Care in Uganda

Very few health centers satisfy the WHO-PEN guidelines for necessary equipment and medications to deliver efficient SCD therapies, notwithstanding inter-facility variation. To appropriately address the rapidly increasing SCD burden in Uganda, improvements in the distribution of human resources, necessary drugs, and technologies, together with the adoption of quality assurance measures, are urgently required. 1968 saw the establishment of Uganda's first and largest dedicated clinic for SCD treatment at Mulago Hospital, which is connected to Makerere University, in Kampala. The SCD clinic is open every day for regular preventative care as well as urgent daytime care services for both adults and children. Pediatricians, nurses, counselors, medical record officers, volunteers, visiting professors, and rotating medical students work in the clinic. Regular visits (every

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one to three months) can be scheduled, notwithstanding there will be significant transportation and other expense problems for families.

The majority of Mulago SCD clinic patients are locals of Kampala and the surrounding districts. Some basic testing is done by an on-site laboratory, and some drugs for routine preventative care are distributed by its pharmacy. Additional laboratory testing, including diagnostic testing for SCD, is paid for by families. Parental counseling opportunities are scarce. In 2010, a thorough clinical database was created for the 11,000 registered patients. From the viewpoint of the personnel, facilities, diagnostic evaluations, staffing, drugs, and counseling are all limited. Healthcare professionals recognized the need for general pediatric staff to expand SCD care and to establish a network of smaller local health facilities and regional centers for SCD clinics. For instance, Nsambya Hospital, a minor hospital in Kampala, runs a weekly SCD clinic and currently employs a steady staff of pediatric physicians and nurses to care for about 100 patients. Good diagnostic capabilities are part of the laboratory's capacity.

2.5.4. Health Knowledge of SCD among Policy Makers

The general knowledge of disabilities and chronic illnesses among policymakers has a big influence on how people are treated, how policies are created, and how they are seen and handled. Sometimes stigma from friends, family, medical professionals, and the general public results from knowledge—or lack thereof. In order to assess the knowledge, attitudes, and self-efficacy of 146 college students majoring in health professions, a 90-item survey was employed. The results showed that students had a poorer (53.9%) than a higher (46.1%) overall grasp of disability (Culp, Rojas-Guyler, Vidourek, & King, 2017). When health professionals have accurate knowledge of disabilities, they feel more competent to care with people who have impairments (Mesa & Tsakanikos, 2014; Lehman, 2009). Most physicians use the knowledge they gained during their residency to manage patients with sickle cell disease (SCD) (Whiteman et al., 2015). However, if medical professionals are not staying up to speed on new research and published works

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about sickle cell disease (SCD), then in certain cases the knowledge they acquired during their medical school may become outmoded. Research has demonstrated that higher levels of comfort are achieved when experience and knowledge are combined. Taking care of individuals with sickle cell disease is made less comfortable when there is insufficient knowledge and expertise. In Nigeria, premarital HIV/AIDS screening is required (Umar & Oche, 2012). Nonetheless, Umar and Oche (2012) found that the nation's religious leaders lack knowledge of HIV/AIDS screening, which negatively impacts their followers. Premarital screening should be expanded to include SCD, according to Arulogun and Adefioye (2010), given the high frequency of SCD in Uganda.

Since the majority of marriage licenses are issued by religious organizations, policymakers are responsible for making sure that prospective spouses go through premarital screening. To increase the program's effectiveness, religious leaders must be included in premarital screening (Dibua, 2010). Despite the high death rate from poor health care, Moronkola and Fadairo (2007) discovered that Ugandans have favorable attitudes and knowledge of sickle cell anemia.

2.5.5. SCD Health policy support and implementation

WHO Recommends that countries with an SCD birth prevalence of 0.5 per 1000 live births should establish separate SCD programs. Organization, 2006 #84, WHO: The focus of strategies and initiatives to reduce morbidity and mortality related to Sickle-Cell Disease (SCD) should be on providing competent care to vulnerable individuals. This strategy offers a range of public health activities to mitigate the effects of sickle cell disease (SCD) in the African Region, including improved understanding, sickness prevention, and early detection. The prevalence, morbidity, and mortality of sickle-cell disease (SCD) can be decreased with the help of interventions like better healthcare delivery, facilities that are clinical, laboratory, diagnostic, and imaging appropriate for different levels of the health system, newborn screening, training for healthcare professionals and the development of

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protocols, genetic counseling and testing, accessibility to healthcare, patient support groups, advocacy, and research. However, the successful implementation of these interventions depends on the commitment of Member States to integrate SCD prevention and control in national health plans and provide an environment conducive for various stakeholders to contribute. Following the Ndeezi et al. 2016 study, Uganda's Ministry of Health implemented a newborn screening program in areas with the greatest sickle cell disease burden, one of the major initiatives done to address the illness's impact.

2.5.6. Health Care Provider Perspectives

Healthcare providers and caregivers must be knowledgeable about treating sickle cell disease (SCD) to provide patients with effective and high-quality treatment. Nurses in particular struggle greatly when taking care of sickle cell disease clients because these patients do not fit the stereotype of the "sick role" that is associated with hospital patients with acute illnesses, according to a study done at Southern University Teaching Hospital in Baton Rouge, Louisiana (Mabien, Labbe, Herbert, & Haynes, 2001). Nurses have been noted to take sickle cell discomfort in patients with sickle cell disease (SCD) less seriously than they do in patients without SCD (Mabien et al., 2001).Healthcare providers have misconceptions about sickle cell disease (SCD) since they don’t know how to treat patients and are afraid of turning SCD patients become junkies from painkillers (Mabien et al., 2001). By failing to provide the necessary analgesics, medical personnel still mistreat SCD patients, especially those who are minorities. Chestnut (1994) looked at the relationship between race and gender in his Service Perception Test (SPT), a pilot research that looked at how several demographics were thought to affect how well patients received medical care. These demographics included age, race, and gender. A survey on patient care at Souther” University Teaching Hospital revealed a deficiency of empathy among the licensed practical nurses. According to this study, SCD patients don’t fully grasp SCD or pain (Mabien, Labbe, Herbert, & Haynes, 2001). Medical professionals were able to express some empathy for patients experiencing suffering from trauma, post-operative

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procedures, and cancer. However, healthcare professionals continued to find it difficult to empathize with patients who had sickle cell disease (SCD) (Mabien et al., 2001).

2.5.7. Stigmatization among SCD patients

According to Weiss, Ramakrishna, and Somma (2006), stigma is "typically characterized by social disqualification of individuals and populations who are identified with particular health problems.". A framework of stigma and discrimination was developed to comprehend stigma connected to health at various socio-ecological levels. As seen in Figure 3 below, it offers a comprehensive perspective on understanding actions to reduce stigma. In 2019 Stangl et al. According to Bulgin, Tanabe, and Jenerette (2018), family, the general public, and healthcare professionals are the main contributors to the stigma associated with health issues.

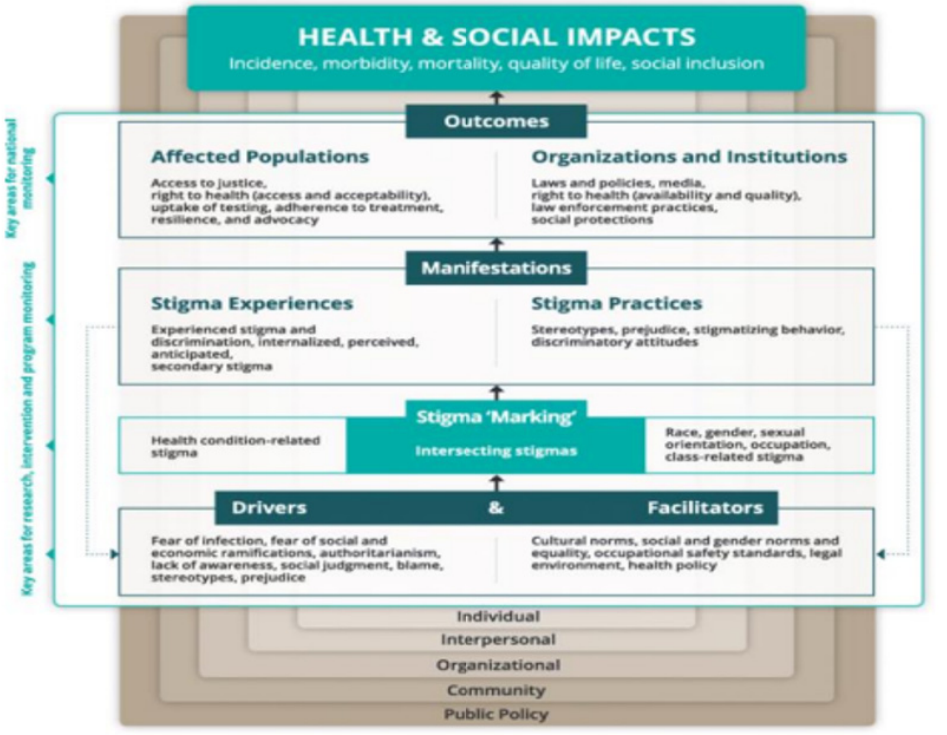

Figure 3. Health, Stigma, and Discrimination Framework

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2.5.8. Myths and Misconceptions about SCD

In Africa and beyond, there are a number of myths and misconceptions that influence people's knowledge and treatment of the disease. In Uganda, for example, children with sickle cell disease (SCD) were believed to have gone through enigmatic cycles of birth, death, and rebirth, according to their cultural beliefs. In certain African societies, following the death of a baby or small kid, family would carry out specific rites and bestow names associated with death from happening again. Furthermore, some literature claims that misbehavior or evil spirits are to blame for Sickle-Cell Disease (SCD). A study from Tanzanian indicates they thought the disease was a god's punishment and that it was more common in the impoverished (Nkingi, 2017). There is a widespread misconception that men do not have the sickle cell gene, according to certain research. Furthermore, Zounon brought up the fact that the general public in Benin is not well informed about the hereditary nature of sickle cell disease (SCD) (Zounon et al., 2012); is not necessarily associated with disease and can be identified through genetic testing. It is not curable by conventional healers. Severe problems with the kidneys, lungs, heart, or brain can arise from sickle cell disease. There is a widespread misconception, nonetheless, that SCD exclusively affects black people. Although anybody can have sickle cell disease (SCD), persons of African, Hispanic-American, Middle Eastern, Indian, and Mediterranean origin are more likely to have it. (Royal et al., 2011).

2.6. Theoretical Framework

The study was grounded within the health belief model (HBM)

2.6.1. Health Belief Model

The goal of the psychological model known as HBM is to forecast behaviors related to preventative healthcare. In 1952, Hochbaum, Rosenstock, and Kegelsin devised the model. The model has been modified to examine a range of health-related activities, including sexual behavior, HIV/AIDS transmission, breast cancer detection, and

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tuberculosis prevention (Abolfotouh et al., 2015). The Health Belief Model states that one's health behavior is influenced by their views as a result of health problems and the importance of taking activities to lessen such threats (Canbulat & Uzun, 2008). If someone believes that they can prevent a bad health condition and believes that following advice will help them avoid the disease, they will do a health-related activity accordingly (Ayele et al., 2012). The HBM accounts for the readiness of individuals to change for a health behavior. Perceived severity is a person's assessment of the ailment's seriousness, whereas perceived susceptibility is a person's conviction that they can get a sickness (Ayele et al., 2012).

Perceived obstacles are related to an individual's judgment on the expenses of the advised action, whereas perceived advantages are concerned with an individual's belief in the effectiveness of recommended activities in lowering the severity or risk of the illness. 1. According to Ayele et al. (2012), the fifth domain, cues to action, stimulates healthy behavior by activating preparedness. The final dimension, self-efficacy, relates to a person's belief in their capacity to behave effectively.

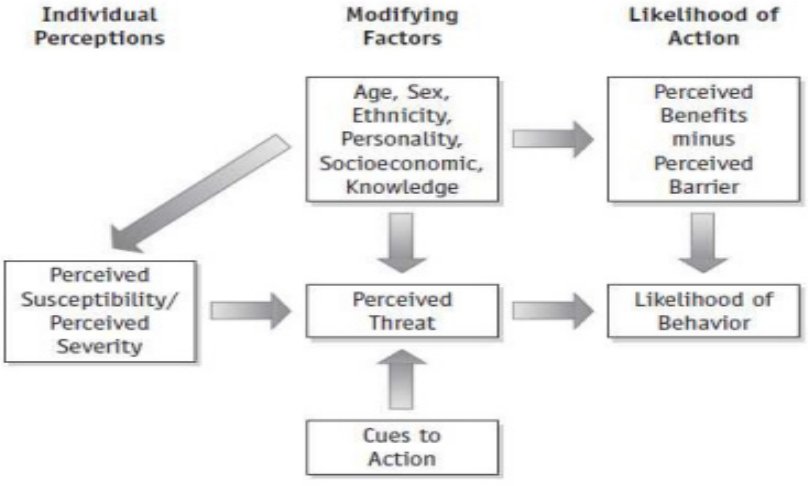

Figure 4. Shows the HBM model derived from "Health behavior theory, research, and practice"

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2.6.2. History of HBM

One of the earliest models of health behavior was created by social psychologists Howard Leventhal, Godfrey M. Hochbaum, S. Stephen Kegeles, and Irwin M. Rosenstock, at the US Public Health Service in the 1950s. Even though mobile X-ray cars visited neighborhoods at a time, researchers and medical professionals were concerned that few individuals were getting screened for tuberculosis (TB). The HBM has been used to predict many different health behaviors, including early diagnosis and being immunized. Understanding vaccination intentions, responses to disease symptoms, adherence to dietary recommendations, lifestyle behaviors (such as sexual risk behaviors), and behaviors associated with chronic illnesses—which may involve both initial behavior change and long-term behavior maintenance—have all been studied using the Health Belief Model (HBM). Even as late as 1988, changes were made to the model to take into account new psychological research regarding the function of self-efficacy in behavior and decision-making.

2.6.3. Theoretical constructs

The theoretical structures of HBM are based on theories from the field of cognitive psychology. It is hoped that adopting a particular health precaution may assist people in avoiding the illness for which they feel they may be at risk.

2.6.4. Applications of the HBM

Initiatives based on the Health Belief Model (HBM) may try to raise perceived seriousness and susceptibility to a health problem by educating people about diseases and their effects, including financial, social, and medical ramifications. Interventions can also aim to alter the cost-benefit analysis of engaging in a health-promoting behavior (i.e., increasing perceived benefits and decreasing perceived barriers) by disseminating information about the efficacy of various behaviors to lower the risk of disease, identifying commonly perceived barriers, offering incentives to engage in health-

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promoting behaviors, and enlisting the support of social networks or other resources. Interventions based on the Health Belief Model (HBM) can also provide cues to action, encouraging and reminding individuals to adopt health-promoting behaviors. Interventions can focus on the societal (e.g., through laws, changes to the physical environment, mass media campaigns) or individual (e.g., working one-on-one with people to encourage involvement in activities important to their health) levels of society. The Health Belief Model has been utilized in numerous research to determine what influences a person's ability to alter behavior as well as their intention to do so. Similar to perceived severity, the respondents expressed little desire to stop their habits because they did not perceive them to have serious consequences. Additionally, there was a weak positive link between the perceived advantages and the respondents' perception that adopting healthy activities would improve their quality of life in general. There was a modest negative association between perceived obstacles and the likelihood of quitting smoking, which meant that the more difficulties that individuals identified with quitting, the less likely they were. The perceived components of the Health Belief Model were correlated with young adult women's intention to quit smoking. The HBM was also employed in a 2016 study (Mo et al., 2016) that looked at the variables linked to physical activity among people with mental illness (PMI) in Hong Kong. Since the HBM model offers a framework for comprehending PMI physical activity levels and is one of the models that is commonly used to describe health behaviors, it was adopted in the study. 443 PMI participated in the study, and their average age was 45. Perceived barriers were revealed to be one of the HBM variables that significantly predicted physical activity. Because it demonstrates how culture can affect the HBM, this study is significant about it. In contrast to American society, Chinese culture places a larger focus on fate and the harmony of the spiritual world than on physical fitness. The HBM does not take into account these extraneous variables, which indicates a flaw in the model and the fact that additional factors besides those included in the model can have an impact on health-related decisions.

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2.6.5. Perceived susceptibility

Arbitrary assessment of the probability of experiencing a health problem. The HBM states that if people think they are predisposed to a certain health issue, they will take actions to lessen their likelihood of experiencing it. Individuals who are thought to be less prone to illness might not think they have a chance of developing that illness. While some may know they could develop the illness, they may believe it is unlikely. If someone believes their chance of being sick is low, they are more likely to engage in dangerous or risky actions. People who think there's a good probability they might personally face a certain health problem are more likely to take precautions against that possibility.

2.6.6. Perceived severity

Refers to an individual's subjective assessment of the gravity of a health problem and any associated adverse effects. The HBM states that when people think a health issue is significant, they are more likely to take action to stop it from getting worse or to lessen its severity. The phrase "perceived severity" describes views on how sickness affects social and professional functioning in addition to opinions regarding the illness itself (e.g., whether it is terminal, may cause disability, or is uncomfortable). For example, someone may think that the flu is not a serious medical condition, but they can think differently if they think that missing a few days of work will have a big financial impact. If people believe they have a high chance of experiencing a certain health condition, they are more inclined to take preventative measures to lower their risk. Researchers found that they could predict whether Australians would get the flu shot by examining their perceived severity of the flu and the number of people who self-reported receiving the influenza vaccine in 2019. To determine perceived severity, they asked, on a scale of 0-10, how severe do you think the flu would be if you got it? They discovered that 31% of respondents believed that getting the flu would be of low severity, 44% believed it would be of moderate severity, and 25% believed it would be of high severity.

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2.6.7. Perceived benefits

Refers to an individual's assessment of the merit or efficacy of engaging in a health-promoting behavior to reduce the risk of disease. It is a crucial element that affects activities related to one's health. A person is more likely to act in a certain way if they believe it would reduce their risk of developing a health problem or diminish the severity of that problem, regardless of the objective facts about how effective the action is.

2.6.8. Perceived threat/barriers

Behaviors connected to health might be affected by barriers to action. The way a person perceives obstacles to changing their behavior is explained by the "perceived barriers" idea. Barriers may keep someone from forming a habit that supports good health even if they think a certain activity will greatly reduce the risk and they regard the health condition as serious. Put another way, for behavior to change, advantages should exceed disadvantages. The inconvenience, cost, risk (like the adverse effects of a medical procedure), and discomfort associated with engaging in the behavior are a few instances of perceived obstacles to action.

2.6.9. Modifying variables

Individual factors can influence how health-related actions are seen, including how serious, vulnerable, advantageous, and challenging they are. These factors include demographic, psychological, and structural factors. There are offered demographic details like age, sex, race, ethnicity, education, and others. Peer and reference group pressure, social class, and personality are only a few examples of psychosocial factors. Prior exposure to a disease and familiarity with a certain disease are just two examples of structural factors. These variables indirectly affect health-related behaviors by shaping how individuals perceive the seriousness, susceptibility, and challenges, as proposed by the HBM.

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2.6.10. Cues to Action

Engaging in behaviors that facilitate health, requires a trigger. Internal signals to act include physiological indicators like pain and suffering. External signals are events that take place or knowledge that people learn from friends, family, the media, or medical professionals that encourage them to practice healthy habits. Triggers to action include things like a dentist's postcard of reminders, media campaigns. Different responses are produced by people based on their perceptions of vulnerability, severity, advantages, and barriers. People who feel they have a significant documented risk of becoming seriously ill.

2.6.11. Self-efficacy

The conviction that one can successfully execute a health activity is known as self-efficacy. Initially, the model was developed to explain participation in health behaviors like receiving a cancer test or a vaccination. Later, more major, long-lasting behaviors including smoking, exercise, and eating were changed using the HBM. The designers of the model recognized the significance of self-efficacy as a key component in altering one's behavior in light of one's health. Schmiege et al., for instance, made a discovery.

2.6.12. Empirical support

The HBM has had a lot of empirical support since the 1950s. It remains one of the best and most studied models for determining causes and predicting health behaviors. An analysis of 18 prospective research and 28 retrospective investigations conducted in 1984 provides insight into the quality of the data supporting each HBM component. The review states that empirical support for the HBM is particularly noteworthy given the variety of populations, health problems, and health-related behaviors reviewed, as well as the numerous study designs and assessment procedures used to test the model. A more recent meta-analysis discovered strong support for the predictive power of perceived

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benefits and barriers, but minimal support for perceived seriousness and susceptibility. Numerous research have provided empirical support from the perspective of chronic illness. The model developed by Becker et al. was used to predict and explain how likely a mother adhere to a diet suggested for her obese children. Interviews with insulin-using diabetics were done by Cerkoney et al. after they attended diabetes education programs at a community hospital. It empirically investigated the connection between the HBM and the level of adherence in type 2 diabetics who experience persistent illness.

2.7. Management of Sickle Cell Disease

Despite improvements in its management, such as better care and transfusion of bone marrow, SCD still results in death rates in Uganda. As a result of its chronic nature, SCD requires long-term medical treatment, which has a negative economic and psychological impact on both the patients and families with SCD (Abioye-Kuteyi et al., 2009). The only known treatment for SCD is bone marrow transplantation, although it is infrequently carried out because of the substantial risks involved (Renaghan et al., 2020) (Oludare & Ogili, 2013). However, identifying carriers during genetic counseling is the greatest strategy for SCD prevention. In Africa, where the disease is high, genetic counseling is cost-effective (HERNANDEZ, 2019). Programs for genetic counseling and premarital screening can lower the incidence of SCD and other hemoglobinopathies in newborns (Shuaibu, 2019). The target population's awareness of SCD, attitude toward genetic testing and counseling, and comprehension of the repercussions of having an afflicted kid are all necessary for the program to be successful (Shuaibu, 2019). Testing for Sickle Cell Disease Genetically to reduce SCD in babies, premarital screening for the diagnosis is essential (Sani & Suleiman, 2014). Premarital screening makes it possible to assess a person's risk of having children due to their health. Premarital genetic screening gives people the opportunity to learn about their genetic susceptibility to disease. Pre-marital testing is thus one of the most effective ways to avoid hereditary illnesses like SCD. The search for SCD carriers and marriages at risk is made easier through screening. Neo-natal

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programs can be adopted to assess the likelihood of fatalities among children with SCD, according to (Sabarense et al., 2015). The type of genotype is probably going to make it easier to diagnose SCD and its problems. To reduce SDC-related mortality, patients' families can be educated. Uganda has launched premarital screening programs, which have proven effective in nations (Ayugi, 2018) like Turkey and Saudi Arabia, these programs are effective in lowering the incidence of hemoglobinopathies (Guler & Karacan, 2007) To persuade prospective spouses to visit genetic counseling clinics before considering marriage, (Hernandez, 2016) stressed the significance of raising awareness and information about the illnesses. It has not been determined how knowledgeable policy-makers are in promoting screening in Uganda.

2.8. Prevention of SCD

Premarital and pre-conception genetic counseling and testing, as well as universal newborn screening, are important methods for preventing SCD. This calls for widespread community education, the provision of free or heavily discounted SCD screening services, and the involvement of important parties like political, religious, and cultural leaders, as well as village health teams. The research as a whole emphasizes the need for policy-makers to have a better understanding, of perceptions, and practices regarding SCD.

Improved financing, resources, policies, and programs, as well as better health outcomes for people with SCD, can all result from greater education and awareness. Newborn examinations according to Bioethics (2001), Newborn screenings are considered the most successful for diagnosing hereditary illnesses. Infections that could be fatal during the first few years of life can be considerably decreased by early detection and treatment of sickle cell disease (SCD) (U.S. Preventive Services Task Force, 2007). Despite the fact that all states now screen for sickle cell disease, it is still less well-known than other blood disorders including hypothyroidism and phenylketonuria (PKU), which are also detected by early screenings (Yang, Andrews, Peterson, Arvind, & Cepeda, 2000).

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III. METHODOLOGY

3.1. Design

A self-examined Google Drive questionnaire was used to conduct a cross-sectional study in Kalangala district, Uganda. A quantitative methodology with the use of a Google form opinionated instrument and a large sample that represented Ugandan policymakers. The methodology facilitated assessing the knowledge level, perception, and practices among policymakers toward SCD and its prevention.

3.2. Research Questions

The research study attempts to answer the following three questions by using an SCD self-examined instrument to survey Ugandan Policy-makers on their existing knowledge, perception, and practices toward sickle cell disease.

- What is the level of knowledge of Policymakers toward SCD in Kalangala?
- What is the practice and perception of policy-makers in Kalangala, related to Sickle Cell Disease?
- What are the recommendations and strategies to improve the knowledge, perception, and practices of policy-makers in Kalangala, Uganda regarding Sickle Cell Disease?

3.3. Participants

The target population of this study was Ugandan policy makers/influencers on the payroll of Kalangala district local Government such as Political leaders (Member of Parliament, District political leaders), Health care professionals, Non-healthcare civil servants, Pensioners, NGO/CSO workers, Media house workers, Religious/Cultural leaders. There were 151 respondents.

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Inclusion and Exclusion Criteria

- The inclusion criteria were as follows:

+ Any staff on the Kalangala district local government payroll

+ Any member of parliament who is a resident of the district

+ Any Non-government organization/NGO staff in the district

+ Any Media house staff who resides in the District

+ Any religious/Cultural leader who resides in the District

- The exclusion criteria were as below.

+ Any staff who absconded from duty

+ Any staff unwilling to participate in the study

+ Any staff who didn't complete the questionnaire.

3.4. Sample Size Estimation

194 respondents were selected from the general population of 684 staff on the Kalangala, using a Standard Formula (https://www.wallstreetmojo.com/sample-size-formula/) to determine the sample size.

$$\text{Sample size, } n = N * \frac{\frac{Z^2 * p * (1-p)}{e^2}}{[N - 1 + \frac{Z^2 * p * (1-p)}{e^2}]}$$

Source (https://cdn.wallstreetmojo.com/wp-content/uploads/2019/06/Sample-Size-Formula.jpg.webp)

Where, the population size is 684, which is denoted by N. The normal distribution's critical value at the necessary degree of confidence (z-score, 1.65) is denoted by Z. p = Sample proportion/standard of deviation (0.5), e = Margin of error (0.05), Sample size, **n**= 194 samples

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3.5. Sampling

This study employed Convenience and Snowball sampling in Kalangala district. The target area is located in southern Central Uganda. The district is coterminous with the Islands in Lake Victoria, the third largest lake in the world. Lake Victoria encircles Kalangala completely and covers 9,066.8 km2. The remaining area, 432.1 km^ (4.8%), is made up of water. The islands that make up the district are referred to as the Ssese Islands.

3.6. Measures

I used HBM to design a few questions on the part of practices and perceptions toward SCD. The study instrument was a self-reported Google form with 20 items, of which 6 were demographic characteristics of the respondents. A questionnaire is a useful tool for conducting quantitative research. With the use of questionnaires, researchers can quickly and affordably gather reliable data (Creswell, 2009). Therefore, using a survey questionnaire that could be used with a large number of participants and duplicated in a different demographic proved to be the most effective way to obtain information from policymakers.

It was also easy for me to disperse questionnaires via social media groups of district staff, NGO, and media workers Watsup and email addresses of particular respondents since Watsup and emailing are the easiest forms of communication in Uganda. I selected survey questionnaires in Google form as the instrument for my study due to their practical nature. I evaluated 151 participants, who were used as a representative of the study population. It was cost-friendly and it gave respondents enough time. Furthermore, it was easy analyse data.

3.7 General Characteristics

The general characteristics of participants consisted of 6 items: age, gender, nature of

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work, marital status, highest level of education, and having biological children.

3.8 Data Collection

A self-examined questionnaire created using Google Drive was used to gather data. The Master of Public Health research conducted in 2011 at the University of Illinois at Urbana-Champaign Graduate College led to the adoption of the data gathering instrument, albeit with some adjustments in the USA, a community survey conducted in 2018 by BMC Public Health among individuals in Kampala, Uganda's Lubaga division and Ugwu 2016; Boadu and Addoah 2018 in Nigeria. The thesis committee members reviewed the questionnaire. A self-examined form generated from Google Drive containing three parts, Part I contained 6 items of demographic information, PART II contained 8 multiple-choice questions testing Knowledge of Sickle cell disease, and PART III contained 6 multiple-choice questions testing the Perception and Practices toward SCD.

3.9 Data Analysis

Data analysis was performed using the Jamovi-2.3.21.0 database and coded using a numerical system. Age was determined by respondents. 14 SCD-related questions were listed on the questionnaire. The score was obtained by considering 50% response. A respondent scored good if more than 3 questions out of the 6 were answered correctly for both knowledge, practices, and perceptions respectively, and was scored poor when 3 and below responses were answered wrongly.

Researchers used frequency tables, and chi-square testing to analyse respondents' sickle cell disease (SCD) knowledge, perception, and practices. To find differences in response rates, $p = 0.05$, the statistically significant alpha p-value, was used. Response rates were grouped according to gender, age, marital status, greatest level of education, and the existence of biological children using the chi-square test.

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3.10 Ethical Considerations

The study was conducted in cooperation with Yonsei University Graduate School of Public Health and the Chief Administrative Officer of Kalangala District local government in Uganda. The data were collected from participants who were willing to complete the Google form. In the survey interviews, no personal identifiers were used. An Excel document containing all of the participant data was obtained. Privacy of respondents was maintained in confidence.

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IV. RESULTS

4.1. Participants recruitment

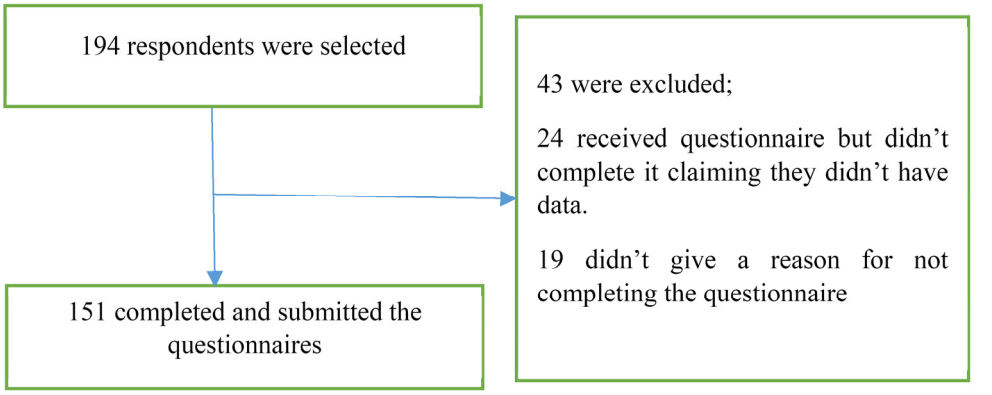

Figure 5. Showing respondents' recruitment process

Out of a total of 194 respondents sampled, 151 completed and submitted the form. A total of 43 respondents were excluded out of these 24 received questionnaires but didn't complete it claiming they didn't have data and 19 respondents didn't give a reason for not completing the questionnaire.

4.2. General Participants Characteristics

A total of 151 policymakers in Kalangala district, Uganda participated in this study. The data collected through the Google form was analyzed using frequency counts. In other words, the responses of each individual were added together to find the highest frequency of occurrence (i.e., the number of times a particular response occurs). Following that, percentage representations of these quantifiable answers to the questions are provided. Categories and general characteristics of the respondents are included as tabulated below in Tables 1 and 2 respectively.

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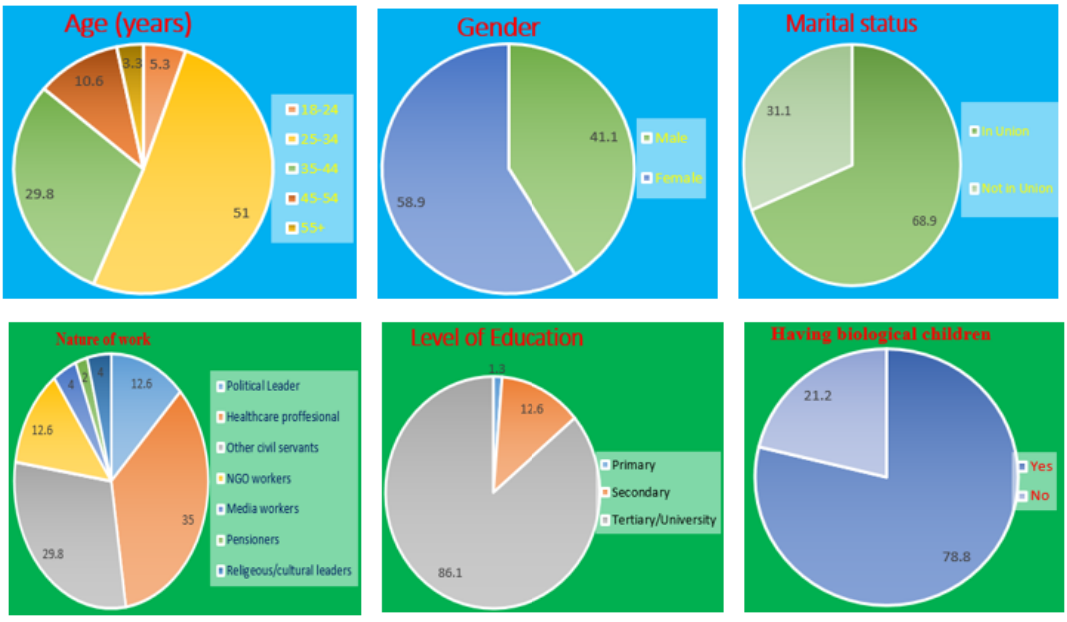

Figure 6. Showing social demographic characteristics of respondents

Figure 6 above reveals that the age group, with the highest percentage (51%) of respondents was between 25-34 years old followed by 35-44 years old (29.8%). Age groups 45-54 and 18-24 years were 10.6% and 5.3% respectively and the smallest age group (3.3%) was 55 years and above. More than half of the total respondents (58.9%) were females compared to their male counterparts (41.1%). Almost three-quarters (68.9%) of the respondents were in Union and less than half (31.1%) of the total respondents were not in Union. In terms of the nature of work, shows that the greater percentage of the respondents were health care professionals with a frequency of 53 and 35% of the total respondents. It indicates that the second largest group of respondents were Non-Health care civil servants with a frequency of 45 and a percentage of 29.8 and this was followed by both NGO/CSO workers and Political leaders which included Members of Parliament, District and sub-county political leaders who ended in equal shares frequency by 19 and 12.6 as a percentage. Media Company workers and Religious/Cultural leaders groups of respondents were the second largest groups by 4%.

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The Pensioners group of respondents was the smallest in number 2 percent. The majority of the respondents (86.1 %) had reached at least a tertiary or university level of education while (12.6 %) reached secondary and only (1.3%) reached primary as their highest level of education. Of the 151 respondents, the majority (78.8%) had biological children, while a minority (21.1%) did not.

4.3. Knowledge of SCD

Table 1. Knowledge of sickle cell disease

Variable	Freq. (N=151)	%
Heard of SCD		
Yes	144	95.4
No	7	4.6
Source of information (n = 144)		
Health care Professional	76	52.8
Friend/family member	57	39.6
TV/Radio	11	7.6
Causes of SCD (n=144)		
Inherited	138	95.8
Don't know	6	4.2
Signs & Symptoms (n=144)		
Yellow eyes	21	14.6
Anemia	44	30.6
Sickle crisis	49	34.0
Don't know	30	20.8
How SCD is Diagnosed (n=144)		
Blood test	97	79.9
Don't know	29	20.1
Medicine for SCD (n=144)		
Conventional medicine	116	80.5
Herbal medicine	1	0.7

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Variable	Freq. (N=151)	%
Don't know	27	18.8
Healthy baby when both parents have SCD (n=144)		
Quarter chance will have the disease	25	17.4
Half chance will have the disease	28	19.4
All children will have the disease	47	32.6
Don't know	44	30.6
Can Health Policy help in SCD		
Yes	132	91.7
No	3	2.1
Don't know	9	6.3

From Table 1 above, the majority (95.4%) of the respondents had ever heard of SCD with the biggest proportion (n=144, 52.8%) hearing about the disease from Health professionals and less than half (39.6%) of the respondents heard it from friends and family compared to those who heard about the disease from TV/Radio (7.6%). According to the survey results, 95.8% of the respondents knew that sickle cell disease (SCD) is an inherited condition. However, a small proportion (4.2%) did not know the cause of the disease. More than three-quarters (79.2%) of the respondents were aware of some signs and symptoms of SCD. Unfortunately, 20.8% of the respondents did not know any signs of the disease. A big percentage (79.9%) of respondents knew how the disease is diagnosed however, still 20.1% didn't know. A big percentage (80.5%) of the respondents knew the medication for SCD as conventional. The results indicate a percentage 0.7% and 1.4% of people consider herbal and prayers respectively as a form of therapy for sickle cell disease while 18.8% do not know the SCD medication. On the question of what is the chance of getting a healthy baby when both parents have the disease, mixed reactions showed up in the responses with 32.6% reporting that all children will have the disease, 19.4% responded that half chance will have the disease and 17.4% mentioned that Quarter chance will have the disease, while 0.7% mentioned that none of the children

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will have the disease. However, more than a quarter (29.9%) of the respondents knew nothing about what chance of having a healthy baby. Responding to the question of Health policy would help in disease prevention and management, the biggest percentage (91.7%) openly accepted with yes and 2.1% said no with a small percentage of 6.3% skeptically answering that they didn't know.

4.4. Practice and perception of sickle cell disease

Table 2. Practice & Perception on sickle cell disease

Variable	Freq. (N)	%
Ever tested for SCD (n=151)		
Yes	33	21.9
No	118	78.1
Reason for testing (n=33)		
Curiosity	13	39.4
Advice from a health worker	20	60.6
Do you know your partner's SCD status (n=151)		
Yes	50	33.1
No	101	66.9
Would you marry someone with SCD?		
Yes	17	11.3
No	118	78.1
Not sure	16	10.6
Can someone with SCD work		
Yes	140	92.7
No	11	7.3
Reason for not working (n=11)		
They are very weak	6	54.5
They fall ill very often	5	45.5

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From Table 2 above, the majority (78.1%) had never tested for SCD only a small percentage 21.9% had ever been screened for SCD and those who tested, had a reason for testing as more than half (60.6%) of these tested as a result of advice from health workers with close to half (39.4%) had curiosity reasons. Unfortunately, a significant proportion (66.9%) of the participants reported that they did not know their partners' sickle cell disease (SCD) status, with only 33.1% being aware of it. Moreover, more than three-quarters (78.1%) of the respondents stated that they would not marry someone with SCD. Just 11.3% of respondents said they would be prepared to marry someone who had sickle cell disease (SCD), and 10.6% said they were not sure. When asked whether someone with SCD can work, the highest percentage (92.7%) accepted yes while (7.3%) of the respondents reported that they cannot work. Among those who responded 'no,' 54.5% cited weakness as the reason while 45.5% reported that they cannot work because they fall ill very often.

4.5. Association between knowledge, practice, perception on sickle cell and relevant factors

Table 3. Association of knowledge score with study variables

Variable		Knowledge Score		*P*-Value
		Good	Poor	
Age	18-34	65(76.5)	20(23.5)	0.707
	35-54	46(75.4)	15(24.6)	
	55 above	3(60)	2(40)	
Gender	Male	48(77.4)	14(22.6)	0.647
	Female	66(74.2)	23(25.8)	
Marital status	In Union	79(76)	25(24)	0.843
	Not in Union	35(74.5)	12(25.5)	
Nature of	Healthcare professionals	48(92.3)	4(7.7)	0.001

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Variable		Knowledge Score		*P*-Value
		Good	Poor	
work	Other Civil servants	49(61.3)	31(38.8)	
	Political leaders	17(89.5)	2(10.5)	
Level of Education	Primary	1(50)	1(50)	0.016
	Secondary	7(36.8)	12(63.2)	
	Tertiary	106(81.5)	24(18.5)	
Having biological children	Yes	90(75.6)	29(24.4)	0.941
	No	24(75)	8(25)	

Table 3 above, shows that out of the 151 participants, 114 (75%) had good knowledge (8–4 points out of 8) about SCD, while the remaining 37 participants, (25%) had poor knowledge. The median score was 4. The age group 18-34years had more respondents and were more knowledgeable about the SCD more than their counter parts of 34-54 years and 55above. The analysis indicates a significant association between the levels of knowledge with the Nature of work with a p-value of 0.001 therefore, there would be a one-in-1,000 chance of observing results at least as extreme. This study rejects the null hypothesis because the null hypothesis is incorrect. The level of education indicates a p-value of 0.016 which is less than the level of significance of 0.05, meaning the result is statistically significant. However, the other study variables of age, gender, marital status, and having biological children showed no significant associations with knowledge

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Table 4. Association of Practices & Perception towards Sickle cell disease with study variables

Variable		Score		*P*-Value
		Good	Poor	
Age	18-34	34(40)	51(60)	0.422
	35-54	31(50.8)	30(49.2)	
	55 above	2(40)	3(60)	
Gender	Male	29(46.8)	33(53.2)	0.620
	Female	38(42.7)	51(57.3)	
Marital status	In Union	48(46.2)	56(53.8)	0.512
	Not in Union	19(40.4)	28(59.6)	
Nature of work	Healthcare professionals	29(55.8)	23(44.2)	0.116
	Other Civil servants	30(37.5)	50(62.5)	
	Political leaders	8(42.1)	11(57.9)	
Level of Education	Primary	2(100)	0(0)	0.075
	Secondary	5(26.3)	14(73.7)	
	Tertiary	60(46.2)	70(53.8)	
Having biological children	Yes	56(47.1)	63(52.9)	0.200
	No	11(34.4)	21(65.6)	

Table 4 above, shows that all the *P*-values are greater than the level of confidence (0.05) therefore, the null hypothesis is retained because the analysis shows no significant association of practice and perception with study variables however, ($P = 0.075$) level of education was very close to 0.05 compared to other variables of age, gender, marital status, nature of work and having biological children.

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V. DISCUSSION

A discussion of the conclusions reached by this investigation is given in this chapter. In Kalangala, Uganda, policymakers were questioned for the study. Male participation in the study was lower than expected; this could be because of their hectic work schedules or unfavorable views about research participation (Francis et al., 2008). The study's findings are consistent with those of Musoke et al. (2014), who discovered that more Ugandan women are using health services. Given that males make the majority of decisions in Uganda, this emphasizes the need of community initiatives.

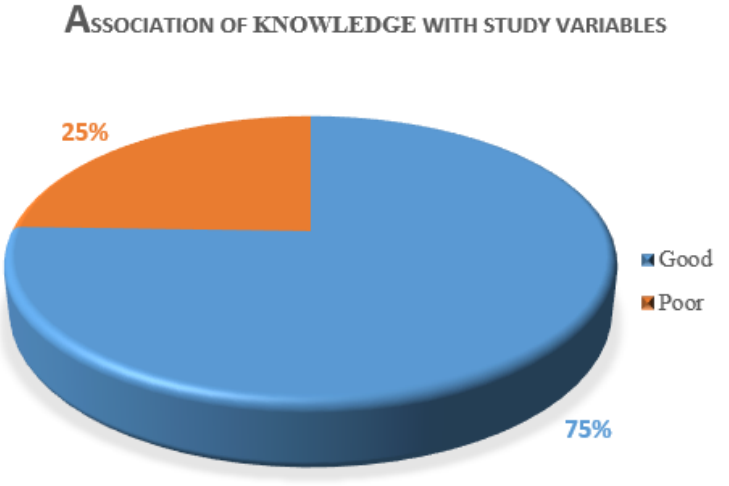

Figure 7. Level of Knowledge of SCD among respondents

From figure 7 above, shows that out of 151 participants, 114 (75%) had good knowledge (8–4 points out of 8), whereas 37 (25%) scored poorly. In Lubaga division, Kampala, Uganda, a study of individuals revealed that 91.2% had heard of sickle cell disease (SCD), with the largest percentage (38.7%) having learned about it via friends and relatives. Although the majority of participants are aware that SCD exists, the fact

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that a significant portion of respondents learned about it from friends or family may indicate that the healthcare system is not making enough of an effort to inform the public about SCD. However, a relatively bigger proportion did not know the signs and symptoms, how SCD is diagnosed and its medication. The results of a study conducted in Uganda suggest that there is a much larger problem that may hinder sickle cell disease (SCD) prevention strategies. Orish et al. 2014 found that schools were the primary source of SCD knowledge in Ghana, while health professionals ranked highest in the study conducted in Uganda. The results also indicate that some individuals consider prayer and herbal medicine as a form of medication for SCD, and some are unaware of whether health policy can influence SCD services, despite being policy influencers in Uganda. In contrast, a different study found that Ugandans living in rural areas thought of prayer as a means of treating chronic illnesses.

These results highlight the need for increased sickle cell disease health education.

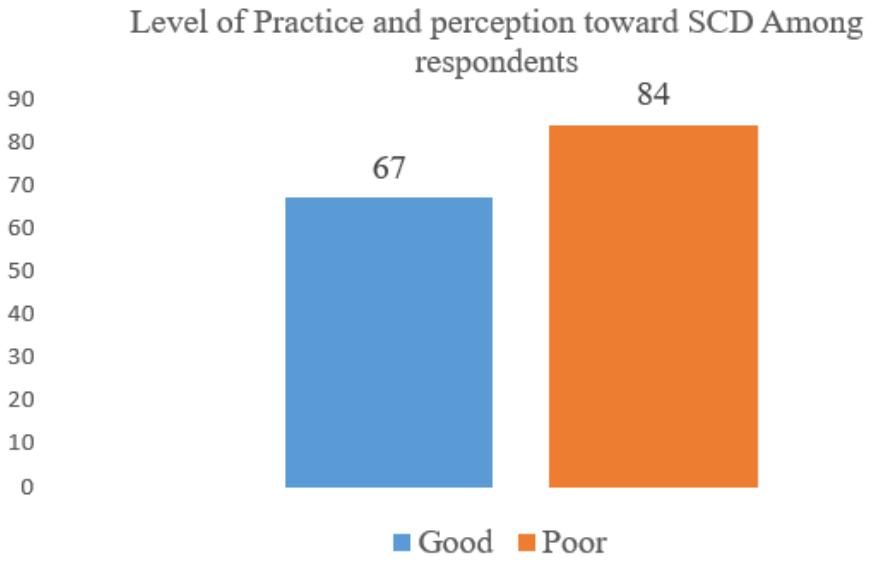

Figure 8. Level of Practice and perception toward SCD among Respondents

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Figure 8 above shows that out of 151 participants, only 67 (44.4%) scored good with practice and perception toward SCD (6–3 points out of 6), and 84 (52.2%) had poor practice and perception toward SCD. The majority of respondents to the poll stated they had never been examined for sickle cell disease (SCD), and a sizable fraction said they had never undergone a sickle cell test. This is due to the fact that the majority of them are misinformed, believe that SCD isn't an emergency, and believe that SCD is a disease that only affects specialist families—not their own—and have restricted access to SCD resources. This suggests a sizable gap in screening services, which could affect family choices and the population's ability to control the disease moving forward. Furthermore, the majority of participants were unaware of their partners' SCD status, which could suggest that screening for SCD is not prioritized before or after marriage but could also have an impact on personal or familial relationships. My study puts more value in addition to the scarce literature focusing on sickle cell disease in Uganda. The study participants were taken from both Government and non-governmental workers, who were representative of the Ugandan policymakers. The study assessed all three aspects of knowledge, practices, and perceptions of an SCD study. There was a wide range of age groups among the participants, providing a good representation of the policymakers. The non-significant *p*-values impacted the study's generalizability. The study entirely relied upon the Primary information provided by participants. Multiple selection questions may have encouraged guessing, which could have introduced bias into the results. Since the selection of the study participants was done using convenient sampling, the rest of the other policymakers in Uganda were not given an equal chance to be represented, so the current results cannot be generalized as representative of the whole Ugandan community. The inadequate data on policymakers toward SCD also limited my study.

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VI. CONCLUSION AND RECOMMENDATIONS

6.1 Conclusion

Notwithstanding the advancements in Ugandan public health policies, the research indicates notable deficiencies in awareness, mindset, and implementation concerning Sickle-Cell Disease (SCD). The study employed convenience and snowball sampling techniques, which are non-probability sampling approaches. According to the survey, the majority of participants had heard of sickle cell disease (SCD), primarily from friends, family, and medical professionals. While most respondents acknowledged that sickle cell disease (SCD) is hereditary, many had never had a test for the condition and stated they would never be able to marry someone who has it, even though many also agreed that health policies can aid in its prevention. The type of work determines the knowledge required, and education level is crucial.

6.2 Recommendations

Conduct similar studies to assess the key drivers of low uptake of SCD services, prevalence, and high unmet needs to inform evidence-based programming. There is a need for targeted programming that is context-specific to address inequities in accessing quality SCD information and services. National sickle cell screening data from Uganda's Central Public Health Laboratories can be used as evidence to design SCD programs. Develop culturally appropriate SBCC that addresses the barriers to uptake and demystifies myths and negative social norms about SCD. Put in place functional multi-sectoral structures to support advocacy at the National & and district levels. Policy and strategy development that encourages male participation in sickle cell disease (SCD) campaigns is essential to raising awareness of the condition and promoting prevention. Furthermore, SCD has to be covered in the community and health center health education

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programs that are currently in place.

6.3 Implications

The study makes room for more research in the future about assessing knowledge, practices, and perceptions toward SCD. Sickle cell anemia is a hematological disorder that is prevalent globally and can be life-threatening. Unfortunately, it is often neglected and there is a high knowledge gap toward SCD in Uganda among both health workers and the general public. Universal newborn screening is considered the best strategy for detecting sickle cell anemia. The seven-nation Consortium on Newborn Screening in Africa for Sickle Cell Disease (CONSA) network of facilities aims to provide early intervention and standardized newborn hemoglobinopathy screening for children with sickle cell disease in sub-Saharan Africa. The overarching hypothesis of CONSA is that, in comparison to past estimates in the area, early infant SCD screening and enrollment in standardized, continuous treatment will lower under-5 mortality. The SCD program is efficient and cost-effective. Living with sickle cell disease (SCD) is causing stigma and discrimination for more people, which lowers quality of life for health-related reasons and may lead to needless stress.

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REFERENCES

Abioye-Kuteyi, E. A., Osakwe, C., Oyegbade, O., & Bello, I. (2009). Sickle cell knowledge, premarital screening, and marital decisions among local government workers in Ile-Ife, Nigeria. *African Journal of Primary Health Care and Family Medicine*, *1*(1), 1-5.

Abolfotouh, M. A., BaniMustafa, A. A. A., Mahfouz, A. A., Al-Assiri, M. H., Al-Juhani, A. F., & Alaskar, A. S. (2015). Using the health belief model to predict breast self-examination among Saudi women. *BMC Public Health*, *15*(1), 1-12.

Ayele, K., Tesfa, B., Abebe, L., Tilahun, T., & Girma, E. (2012). Self-care behavior among patients with diabetes in Harari, Eastern Ethiopia: the health belief model perspective. *PLoS One*, *7*(4), e35515.

Ayugi, B. (2018). *Knowledge, attitude, and practices of health workers towards sickle cell screening in Tororo District, Uganda* Makerere University].

Boadu, I. (2018). AddoahT (2018) Knowledge, Beliefs and Attitude towards Sickle Cell Disease among University Students. *J Community Med Health Educ*, *8*(593), 2161-0711.1000593.

Boyd, J. H., Watkins, A. R., Price, C. L., Fleming, F., & DeBaun, M. R. (2005). Inadequate community knowledge about sickle cell disease among African-American women. *Journal of the National Medical Association*, *97*(1), 62.

Canbulat, N., & Uzun, Ö. (2008). Health beliefs and breast cancer screening behaviors among female health workers in Turkey. *European Journal of Oncology Nursing*, *12*(2), 148-156.

Carden, M. A., & Little, J. (2019). Emerging disease-modifying therapies for sickle cell disease. *Haematologica*, *104*(9), 1710.

Creary, M., Williamson, D., & Kulkarni, R. (2007). Sickle cell disease: current activities, public health implications, and future directions. *Journal of women's health*, *16*(5), 575-582.

Dennis-Antwi, J. A., Ohene-Frempong, K., Anie, K. A., Dzikunu, H., Agyare, V. A., Okyere Boadu, R., Sarfo Antwi, J., Asafo, M. K., Anim-Boamah, O., & Asubonteng, A. K. (2019). Relation between religious perspectives and views on sickle cell disease research and associated public health interventions in Ghana. *Journal of genetic counseling*, *28*(1), 102-118.

Diop, S., & Pirenne, F. (2021). Transfusion and sickle cell anemia in Africa. *Transfusion Clinique et Biologique*, *28*(2), 143-145.

Guler, E., & Karacan, M. (2007). Prevalence of beta-thalassemia and sickle cell anemia trait in premarital screening in Konya urban area, Turkey. *Journal of pediatric hematology/oncology*, *29*(11), 783-785.

Hernandez, A. G. (2016). *Effective strategies used to describe and address the burden of sickle cell*

연세대학교
YONSEI UNIVERSITY

disease in the Republic of Uganda: The Uganda Sickle Surveillance Study University of Cincinnati].

HERNANDEZ, A. G. (2019). EPIDEMIOLOGICAL EVALUATION OF THE NATIONAL SICKLE CELL SCREENING PROGRAM IN THE REPUBLIC OF UGANDA.

Horiuchi, S. S., Zhou, M., Snyder, A., & Paulukonis, S. T. (2022). Hematologist encounters among Medicaid patients who have sickle cell disease. *Blood Advances*, *6*(17), 5128-5131.

Howard, J., Hart, N., Roberts-Harewood, M., Cummins, M., Awogbade, M., Davis, B., & Committee, B. (2015). Guideline on the management of acute chest syndrome in sickle cell disease. *British journal of hematology*, *169*(4), 492-505.

Kambasu, D. M., Rujumba, J., Lekuya, H. M., Munube, D., & Mupere, E. (2019). Health-related quality of life of adolescents with sickle cell disease in sub-Saharan Africa: a cross-sectional study. *BMC hematology*, *19*(1), 1-9.

Katz, M. L., Smith-Whitley, K., Ruzek, S. B., & Ohene-Frempong, K. (2002). Knowledge of stroke risk, signs of stroke, and the need for stroke education among children with sickle cell disease and their caregivers. *Ethnicity and Health*, *7*(2), 115-123.

Leibovitch, J. N., Tambe, A. V., Cimpeanu, E., Poplawska, M., Jafri, F., Dutta, D., & Lim, S. H. (2022). l-glutamine, crizanlizumab, voxelotor, and cell-based therapy for adult sickle cell disease: Hype or hope? *Blood Reviews*, *53*, 100925.

Martinez, R. M., Osei-Anto, H. A., & McCormick, M. (2020). Addressing Sickle Cell Disease: A Strategic Plan and Blueprint for Action.

Nadel, M. V. (1975). The hidden dimension of public policy: private governments and the policy-making process. *The Journal of Politics*, *37*(1), 2-34.

NAMAZZI, R., CONROY, A., BOND, C., GOINGS, M., DATTA, D., WARE, R., OPOKA, R., & JOHN, C. (2022). PI-08: HIGH DISEASE BURDEN, MORBIDITY, AND MORTALITY AMONG CHILDREN WITH SICKLE CELL ANAEMIA IN UGANDA. *HemaSphere*, *6*, 13.

Natukunda, B., Schonewille, H., Ndugwa, C., & Brand, A. (2010). Red blood cell alloimmunization in sickle cell disease patients in Uganda. *Transfusion*, *50*(1), 20-25.

Ndeezi, G., Kiyaga, C., Hernandez, A. G., Munube, D., Howard, T. A., Ssewanyana, I., Nsungwa, J., Kiguli, S., Ndugwa, C. M., & Ware, R. E. (2016). Burden of sickle cell trait and disease in the Uganda Sickle Surveillance Study (US3): a cross-sectional study. *The Lancet Global Health*, *4*(3), e195-e200.

Nkingi, R. C. (2017). *Experiences of Living with Sickle Cell Disease among Adults Attending Clinic at Muhimbili National Hospital, Dar-Es-Salaam, Tanzania* Muhimbili University of Health and Allied Sciences].

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Nnebe-Agumadu, U., Adebayo, I., Erigbuem, I., James, E., Kumode, E., Nnodu, O., & Adekile, A. (2021). Hydroxyurea in children with sickle cell disease in a resource-poor setting: Monitoring and effects of therapy. A practical perspective. *Pediatric Blood & Cancer*, *68*(6), e28969.

Ogamdi, S. O., & Onwe, F. (2000). A pilot study comparing the level of sickle cell disease knowledge in a university in southeastern Texas and a university in Enugu, Enugu State, Nigeria, West Africa. *Ethnicity & Disease*, *10*(2), 232-236.

Olatona, F., Odeyemi, K., Onajole, A., & Asuzu, M. (2012). Effects of health education on knowledge and attitude of youth corps members to sickle cell disease and its screening in Lagos State.

Oludare, G. O., & Ogili, M. C. (2013). Knowledge, attitude, and practice of premarital counseling for sickle cell disease among youth in Yaba, Nigeria. *African journal of reproductive health*, *17*(4).

Osbourne, C. (2012). *Sickle cell disease awareness amongst college students* University of Illinois at Urbana-Champaign].

Phillips, S., Chen, Y., Masese, R., Noisette, L., Jordan, K., Jacobs, S., Hsu, L. L., Melvin, C. L., Treadwell, M., & Shah, N. (2022). Perspectives of individuals with sickle cell disease on barriers to care. *PLoS One*, *17*(3), e0265342.

Rantanen, E., Hietala, M., Kristoffersson, U., Nippert, I., Schmidtke, J., Sequeiros, J., & Kääriäinen, H. (2008). What is ideal genetic counseling? A survey of current international guidelines. *European Journal of Human Genetics*, *16*(4), 445-452.

Reid, C., & Rodgers, G. (2007). Sickle cell disease: demystifying the beginnings. *Renaissance of sickle cell disease research in the genomic era. London, Great Britain: World Scientific*, 1-12.

Renaghan, A. D., Jaimes, E. A., Malyszko, J., Perazella, M. A., Sprangers, B., & Rosner, M. H. (2020). Acute kidney injury and CKD associated with hematopoietic stem cell transplantation. *Clinical Journal of the American Society of Nephrology: CJASN*, *15*(2), 289.

Royal, C. D., Jonassaint, C. R., Jonassaint, J. C., & De Castro, L. M. (2011). Living with sickle cell disease: traversing 'race and identity. *Ethnicity & Health*, *16*(4-5), 389-404.

Sabarense, A. P., Lima, G. O., Silva, L. M. L., & Viana, M. B. (2015). Survival of children with sickle cell disease in the comprehensive newborn screening program in Minas Gerais, Brazil. *Pediatrics and International Child Health*, *35*(4), 329-332.

Sani, A. M., & Suleiman, F. (2014). KNOWLEDGE, ATTITUDE, AND PRACTICE OF PRE-MARITAL GENETIC COUNSELLING AND TESTING OF SICKLE CELL DISEASE AMONG WOMEN IN ZARIA, NIGERIA. *West African Journal of Nursing*, *25*(2).

Shuaibu, M. S. (2019). Knowledge, beliefs, and attitude of students at Kampala International

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University towards sickle cell disease.

Stewart, K. A. (2007). *An examination of African American college students' knowledge and attitudes regarding sickle cell disease and sickle cell disease carrier testing: A mixed methods study*. The University of Alabama at Birmingham.

Thomas, P. W., Higgs, D. R., & Serjeant, G. R. (1997). Benign clinical course in homozygous sickle cell disease: a search for predictors. *Journal of Clinical Epidemiology*, *50*(2), 121-126.

Thomson, A. M., McHugh, T. A., Oron, A. P., Teply, C., Lonberg, N., Tella, V. V., Wilner, L. B., Fuller, K., Hagins, H., & Aboagye, R. G. (2023). Global, regional, and national prevalence and mortality burden of sickle cell disease, 2000–2021: a systematic analysis from the Global Burden of Disease Study 2021. *The Lancet Haematology*.

Treadwell, M. J., McClough, L., & Vichinsky, E. (2006). Using qualitative and quantitative strategies to evaluate knowledge and perceptions about sickle cell disease and sickle cell trait. *Journal of the National Medical Association*, *98*(5), 704.

Tusuubira, S. K., Nakayinga, R., Mwambi, B., Odda, J., Kiconco, S., & Komuhangi, A. (2018). Knowledge, perception, and practices towards sickle cell disease: a community survey among adults in Lubaga division, Kampala Uganda. *BMC Public Health*, *18*(1), 1-5.

Yousef, A. A., Shash, H. A., Almajid, A. N., Binammar, A. A., Almusabeh, H. A., Alshaqaq, H. M., Al-Qahtani, M. H., & Albuali, W. H. (2022). Acute chest syndrome in pediatric sickle cell disease: a 19-year tertiary center experience. *Annals of Thoracic Medicine*, *17*(4), 199.

Zounon, O., Anani, L., Latoundji, S., Sorum, P. C., & Mullet, E. (2012). Misconceptions about sickle cell disease (SCD) among lay people in Benin. *Preventive medicine*, *55*(3), 251-253.

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Appendix 1. Letter of Cooperation from Yonsei University

연세대학교 보건대학원
GRADUATE SCHOOL OF PUBLIC HEALTH, YONSEI UNIVERSITY

서울특별시 서대문구 성산로 250(신촌동 134)
전화 : (02) 2228-1505~6
팩스 : (02) 392-7734

250 Seungsan-no, Seodaemun-gu, Seoul, Korea
TEL : + 82 2 2228-1505~6
FAX : + 82 2 392-7734

24 July 2023

The Chief Administrative Officer
Kalangala District Local Government
P.O BOX 2 Kalangala, Uganda

Through,
The District Health Officer
Kalangala District Local Government

SUBJECT: REQUEST FOR AUTHORITY TO COLLECT PRIMARY DATA FROM KALANGALA DISTRICT LOCAL GOV'T STAFF FOR THE PURPOSE OF THESIS WRITING:

This letter serves to inform you that Anthony Byarugaba is a bona fide student pursuing a Master's degree in Global Health Policy and Financing at the Graduate School of Public Health, Yonsei University, South Korea, for the period August 2022 to February 2024. Students are requested to write a thesis as the fulfillment of the Master's degree program. His area of interest is Accessing knowledge, perceptions, and practices on Sickle Cell Disease among policy-makers and influencers in Uganda, Kalangala study area. Among the respondents include; political leaders (Members of Parliament, District & sub-county political leaders), Health care professionals (DHMT, Health workers), other civil servants (Non-Health care professionals), NGO/CSO workers, Media company workers, Pensioners, and religious and cultural leaders.

To assist his academic accomplishment, we kindly request that you allow him to interview your staff through an online survey using a Google form. This self-administered questionnaire has been designed for data collection for this thesis, and their response will be very imperative and exclusively used for this study in Uganda's evidence-based approach to fighting infectious diseases.

Thanks for your support in advance, and please feel free to contact us if you need further information.

Respectfully,

Whiejong M. Han, Ph.D.
Chair, Department of Global Health and Disease Control
Graduate School of Public Health
Yonsei University, Seoul, Korea

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Appendix 2. Letter of Cooperation from Kalangala District Local Government

THE REPUBLIC OF UGANDA

KALANGALA DISTRICT LOCAL GOVERNMENT
Office of the Chief Administrative Officer

P.O Box 2, Kalangala, Uganda
Tel: 0772873729/0701873729 **27th July 2023**

Ref: In correspondence to this
Subject please quote: **CR/219/3**
Your Ref:

The Chair, Department of Global Health and Disease Control
Graduate School of Public Health
Yonsei University, Seoul, Korea

Dear Sir/Madam;
Re: Request for Authority to Collect Primary Data from Kalangala District Local Gov't Staff for the Purpose of Thesis Writing

I refer to your communication to my office, dated 24th July 2023 as per the above mentioned subject matter.

This communication thus, serves to allow your student, Anthony Byarugaba to interview the staff of Kalangala District Local Government through an online survey using a google form as part of his thesis for award of the Master's Degree in Global Health Policy and Financing of the Graduate School of Public Health of Yonsei University, South Korea.

I also take this opportunity to thank the Korean Government for Sponsoring him to undertake his studies at you prestigious University and we wish him success in his studies
Thank you;

KALANGALA DISTRICT LOCAL GOVERNMENT
27 JUL 2023
P. O. BOX 2, KALANGALA
CHIEF ADMINISTRATIVE OFFICER

.....
KYOMYA FRIDAY
CHIEF ADMINISTRATIVE OFFICER

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Appendix 3. Survey questionnaire

PART 1: Demography:			
Variable	**Respondent's Category**		
Age (Years)	A. 18–24 B. 25-34 C. 35-44 D. 45-54 E. 55 Above		
Gender	A. Male	B. Female	C. Prefer not to say
What is the nature of your work?	A. Political leader (Member of Parliament, District & sub-county political leaders) B. Health care professional (DHMT, Health workers) C. Other civil servants (Non-Health care professional) D. NGO/CSO worker E. Media company worker F. Pensioner G. Religious/Cultural leader		
Marital status	A. In union	B. Not in union	
What is your highest level of Education?	A. Primary	B. Secondary	C. Tertiary/University
Do you have biological children	A. Yes	B. No	

No.	**Question**	**Response**
PART II: Knowledge of Sickle Cell Disease		
1	Have you ever heard of sickle cell disease?	A. Yes B. No
2	Where did you get the information about sickle cell disease?	A. Health Professional B. Internet C. Friend/family D. TV/Radio E. Other specify….

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3	How is Sickle cell disease transmitted?	A. Inherited B. Sexually transmitted disease C. Don't know
4	Mention any signs and symptoms of sickle cell disease	A. Yellow eyes B. Yellow skin C. Anemia D. Pain crisis or sickle crisis E. Priapism F. Don't know
5	How is Sickle Cell Disease tested and diagnosed?	A. Blood test B. Genetic test C. Urine test D. Don't know
6	What Medication is for people living with sickle cell disease?	A. Conventional medicine by Health professionals B. Herbal medicine C. Prayers D. Don't know
7	Is there a chance of getting a healthy baby when both parents have Sickle Cell Disease?	A. A quarter (1/4) of the children will have the disease B. Half (1/2) of the children will have the disease C. All the children will have the disease D. None of the children will have the disease E. Don't know
8	Do you think **health policy** can help people with sickle cell disease and their caregivers?	1. Yes 2. No 3. Don't Know
PART III: Perception and Practices towards Sickle Cell Disease		
9	Have you ever tested for Sickle Cell Disease?	A. Yes B. No
10	If yes, why did you test for Sickle Cell Disease?	Short answer…….
11	Do you know your partner's SCD status?	A. Yes B. No

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12	Would you marry someone with SCD?	A. Yes B. No C. Don't know
13	Can someone with Sickle Cell Disease work?	A. Yes B. No
14	If No? Why do you think so?	……..
Thank you for your time and for participating in this study		