The Relationship Between Support Systems and Disease Burden for Families Coping with Sickle Cell Disease in South Africa and Cameroon

by

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Thesis submitted in partial fulfillment of
the requirements for the degree of
Master of Science in the Duke Global Health Institute
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ABSTRACT

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Abstract

Background: Sickle cell disease (SCD) is a debilitating genetic blood disorder that seriously impacts the quality of life of affected individuals and their families. With 85% of cases occurring in sub-Saharan Africa, it is essential to identify the barriers and facilitators of optimal outcomes for people with SCD in this setting. This study focuses on understanding the relationship between support systems and disease outcomes for SCD patients and their families in Cameroon and South Africa.

Methods: This mixed-methods study utilizes surveys and semi-structured interviews to assess the experiences of 29 SCD patients and 28 caregivers of people with SCD across three cities in two African countries: Cape Town, South Africa; Yaoundé, Cameroon; and Limbe, Cameroon.

Results: Patients in Cameroon had less treatment options, a higher frequency of pain crises, and a higher incidence of malaria than patients in South Africa. Social support networks in Cameroon consisted of both family and friends and provided emotional, financial, and physical assistance during pain crises and hospital admissions. In South Africa, patients relied on a strong medical support system and social support primarily from close family members; they were also diagnosed later in life than those in Cameroon.

Conclusions: The strength of medical support systems influences the reliance of SCD patients and their caregivers on social support systems. In Cameroon the health care system does not adequately address all factors of SCD treatment and social networks of family and friends are used to complement the care received. In South Africa, strong medical and social support systems positively affect SCD disease burden for patients and their caregivers. SCD awareness campaigns are necessary to reduce the incidence of SCD and create stronger social support networks through increased community understanding and decreased stigma.

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1. Introduction

1.1 Background

Sickle cell disease (SCD) is a group of genetic blood disorders characterized by the presence of sickle-shaped red bloods cells. These misshapen red blood cells form due to the production of sickle hemoglobin (HbS). HbS results when a single nucleotide substitution (Thymine for Adenine; G**A**G → G**T**G) in the beta-globin subunit of the hemoglobin molecule produces valine instead of glutamic acid. This hemoglobin variant is found on chromosome 11 (Davies & Brozovic, 1989). HbS polymerizes in low-oxygen conditions, altering the structure of red blood cells from their typical biconcave shape into rigid crescents with a reduced life-span (Kanter & Kruse-Jarres, 2013). These sickled red blood cells can easily stick together and block blood vessels leading to severe pain and damage to the body's tissue and vital organs (D. C. Rees, Williams, & Gladwin, 2010). Having one copy of HbS and one normal copy of the gene (HbA) results in a heterozygous genotype (HbAS) where the individual is considered a carrier of the sickle cell trait. Those with HbAS will produce both HbA and HbS and often do not display symptoms of SCD unless in rare cases of severe dehydration or strenuous physical activity (Tantawy, 2014). As a result, many of the individuals with HbAS, especially those living in areas without access to newborn screening, are unaware that they possess the trait (Gallo et al., 2010). SCD is a Mendelian disease inherited in an autosomal

recessive pattern. A person that inherits HbS from both parents is considered to have sickle cell anemia (HbSS).

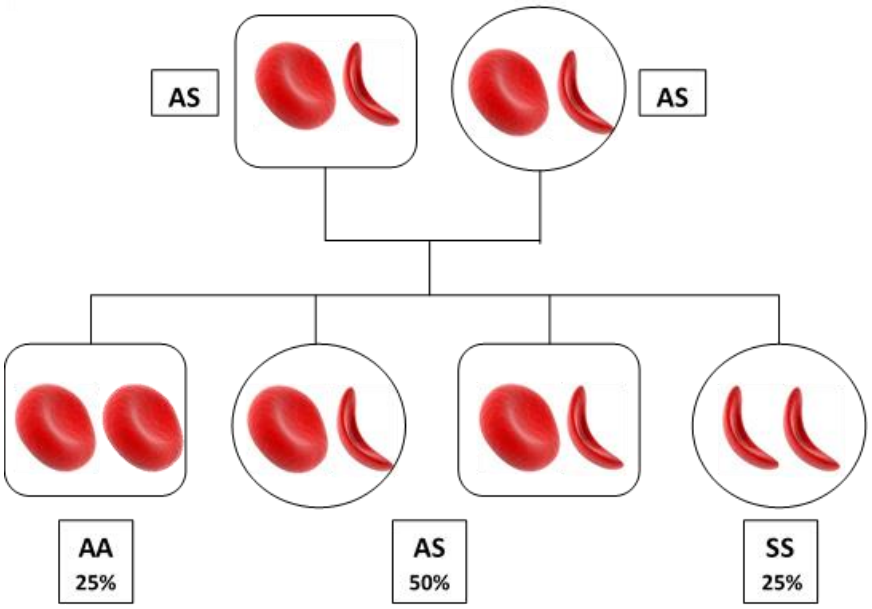

Figure 1 Sickle cell disease inheritance pattern for two heterozygous sickle cell trait carriers

As seen in **Figure 1**, for each child conceived by two sickle cell carriers there will be a 25% chance of a normal genotype (HbAA), a 50% chance of a heterozygous HbAS genotype, and a 25% chance of a homozygous SS genotype which results in sickle cell anemia (Wilkie, Johnson, Mack, Labotka, & Molokie, 2010). Other forms of SCD are inherited in the same pattern and result from combinations of HbS with other abnormal hemoglobin variants. These include hemoglobin SC disease and hemoglobin Sβ thalassemia. However, HbSS is the most common form of SCD and is considered the most severe (Kanter & Kruse-Jarres, 2013).

The first published case of SCD, describing a patient with crescent shaped blood cells and severe anemia, was led by Dr. James Herrick in 1910 (Herrick, 2001). Nearly forty years later the condition was described by Pauling and Itano as the first "molecular disease" after they identified differences between HbA and HbS through electrophoresis (Pauling, Itano, & et al., 1949). Despite the early genetic understanding of SCD there is still relatively little understanding of the large range in phenotypic presentation (Adekile, 2005; Muszlak et al., 2015). There has also been limited progress in the treatment of SCD; despite its discovery being over a century ago, there is only one drug, hydroxyurea, approved to treat the broad range of SCD symptoms (Heeney & Ware, 2008). The slow progression in the understanding and treatment of this disease may be caused in part by the lack of attention SCD receives. SCD is a disease that many consider to be neglected and critically underfunded (Ware, 2013). The absence of adequate resources and awareness for this disease has devastating consequences for both prevention and treatment capacities. Because SCD is a chronic condition, patients require comprehensive, lifelong care that often spans many sectors of the healthcare system (Kanter & Kruse-Jarres, 2013).

1.2 SCD Related Health Concerns

People with SCD are afflicted with a variety of morbidities affecting all systems of the body. Symptoms often begin at around 6 months old as the fetal hemoglobin (HbF) levels in the blood begin to decline (Kanter & Kruse-Jarres, 2013). While some of

the more prominent health concerns (pain, acute chest syndrome, stroke, and infection) are detailed below, the complete scope of multi-organ complications is extensive.

1.2.1 Pain

The hallmark symptom of SCD is pain. Those affected by SCD have periodic episodes of severe pain which are commonly referred to as acute vaso-occlusive crises (VOC) or vaso-occlusive episodes (VOE) (Alli et al., 2014; Kanter & Kruse-Jarres, 2013). VOEs are the most common cause of hospital admission for SCD patients (Ballas & Lusardi, 2005). A high number of VOEs per year is a measure of clinical severity and has been shown to predict early mortality (Platt et al., 1991). VOEs occur when the sticky, rigid sickled-cells in a patient with HbSS cannot pass through blood vessels that are only designed to accommodate typical round, pliable red blood cells. The misshapen cells can form adhesions with the endothelium of blood vessels and block blood flow, resulting in severe pain (Ndefo, Maxwell, Nguyen, & Chiobi, 2008). These blockages can occur throughout the body in areas such as bones, muscles, and tissue. VOEs can be triggered by a variety of factors that vary between patients. Common causes of VOEs include dehydration, strenuous physical activity, folate deficiency, exposure to cold, infection or illness, stress, and anxiety (Alli et al., 2014). While still unpredictable overall, VOEs can be largely avoided with proper care and avoidance of triggers (Ansong, Akoto, Ocloo, & Ohene-Frempong, 2013). Aside from causing intense pain, a VOE also damages the affected tissue through the depravation of blood and oxygen followed by the sudden

restoration of blood flow; the cycle between ischemia and reperfusion causes oxidative and inflammatory stress to the tissue that results in damage over time (D. C. Rees et al., 2010).

In addition to the acute pain brought on by a VOE, those with SCD often suffer from various sources of chronic pain as well. Complications such as leg ulcers, avascular necrosis, and tissue damage are all sources of long-term, debilitating pain (Wilkie et al., 2010). Hip necrosis is the most common orthopedic complication and can progress from limping and discomfort to an inability to walk (Ansong et al., 2013). The knee is the second most common site of necrosis followed by small joints in the hands and feet (Alli et al., 2014). Damage to organs over time can cause organ failure which is another source of chronic pain; heart disease and renal disease are both common occurrences in patients with SCD (D. C. Rees et al., 2010). The pathophysiology of pain in SCD varies widely and some patients also report neuropathic pain and sensitivity to touch (Chakravorty & Williams, 2015).

1.2.2 Acute chest syndrome

Acute chest syndrome (ACS) is a life-threatening complication of SCD that affects the respiratory system. It is the second most common cause of hospitalization for SCD patients, behind VOEs. It is the leading cause of mortality for SCD patients, accounting for about 25% of all SCD related deaths (Vichinsky et al., 1997). Repeated instances of ACS can damage the lungs over time and can lead to decreased lung

function and chronic lung diseases. It manifests more severely in adults over the age of twenty and is often triggered by a VOE. Severe ACS cases can lead to respiratory failure (Vichinsky et al., 2000). ACS is often more mild in children than adults and occurrence in children is often linked to infection (Vichinsky et al., 1997). Symptoms for ACS include severe pain, fever, and shortness of breath. It is often misdiagnosed as a typical VOE; ACS can also develop during a VOE, sometimes after it has already been tested for and ruled out. Because an x-ray is required for diagnosis, patients in resource-limited settings may not have the means for testing (Nansseu et al., 2015).

1.2.3 Stroke

Strokes are a major source of morbidity and mortality in people with SCD and are caused when the blood supply to the brain is interrupted. The depravation of oxygen to the brain can lead to severe neurological damage and death. In SCD patients, strokes occur most often in between 2-11 years of age and affect up to 30% of SCD patients (Alli et al., 2014). Ischemic strokes are the most common type of stroke, making up 87% of the strokes in those affected by SCD. They occur when the blood supply to the brain is blocked such as in cases of thrombosis, embolism, or ischemia. Hemorrhagic strokes are the result of a ruptured blood vessel. While they are more rare than Ischemic strokes, they have a higher mortality rate (Menaa, 2013). A silent cerebral infarct (SCI) is another type of stroke that does not exhibit symptoms. Damage from SCIs can be picked up on

MRIs and transcranial dopplers. They are more common in children and occur in 27% of those with HbSS before age 6 and 37% by age 14 (DeBaun et al., 2012).

1.2.4 Infection

SCD patients are highly susceptible to infections with children being at the greatest risk; infection is the leading cause of mortality in pediatric SCD patients. Their increased susceptibility to infection is often linked to poor splenic function. The spleen functions as a filter to remove old and damaged cells and also produces antibodies; when it is damaged through overuse, scarring, and crises, the immune system is compromised (Booth, Inusa, & Obaro, 2010; Kanter & Kruse-Jarres, 2013). Decreased splenic function leaves patients more susceptible to the risk of life-threatening illness from encapsulated bacteria and malaria (Alli et al., 2014). Those with SCD are predisposed to other infections as well including *Escherichia coli*, urinary tract infection, pneumonia and other respiratory illnesses, dental infections, and cholecystitis (Booth et al., 2010). Frequent blood transfusions increase the risk of Hepatitis B and C and a high red blood cell turnover increases the risk of parvovirus (Booth et al., 2010). Other problems related to SCD such as zinc deficiency, iron overload, malnutrition, and the presence of tissue necrosis increase the risk of infection as well (Alli et al., 2014). In addition to the dangers of an infection itself, infections can also cause other SCD events such as VOEs, ACS, sequestration episodes, and hyperhemolysis syndrome (Ansong et al., 2013).

1.3 SCD Treatments

SCD patients can suffer from a multitude of health problems; therefore, they require many treatments specific to the current medical concerns they are facing. For example, a patient in crisis may require antibiotics, intravenous hydration, and strong pain medication. To temper the effects of chronic pain, pain medications of varying intensity are often necessary on a daily basis as well. However, few treatments are available that actually prevent or treat the overall effects of SCD. Hydroxyurea, blood transfusions, nutritional supplements, and preventative measures are detailed below as treatment strategies that improve a SCD patient's health on a long-term basis.

1.3.1 Hydroxyurea

Hydroxyurea, or hydroxycarbamide, is a form of chemotherapy that is the sole FDA approved drug available to lessen SCD severity. It functions by increasing the production of fetal hemoglobin, as well as decreasing the production of leukocytes and reticulocytes. Hydroxyurea has been shown to reduce the number of VOEs and hospitalizations in SCD patients (Lebensburger et al., 2015). Studies have shown that pediatric patients taking the maximum tolerated dose of hydroxyurea over several years showed significant reductions in organ damage, chronic hypoxemia, and stroke, as well as fewer VOEs and hospitalizations (Kanter & Kruse-Jarres, 2013). It requires a once daily dose and is effective in both children and adults. While limited data exist on the health effects of prolonged use, current studies have not validated any concerns of long-

term toxicity and instead show reductions in overall morbidity and mortality (Heeney & Ware, 2008). Despite its proven effectiveness, the prescription of hydroxyurea varies widely between providers and facilities (A. L. Rees, 2015). In high-resource settings hydroxyurea may be underutilized due to providers' uncertainty about its long-term effects; in low-resource settings it is often unavailable due to financial restraints or the lack of laboratory infrastructure necessary to frequently test a patient's blood levels (Kanter & Kruse-Jarres, 2013).

1.3.2 Blood transfusions

Blood transfusions have been shown to reduce the frequency of VOE, ACS, and stroke by reducing the amount of HbS in the blood (Kanter & Kruse-Jarres, 2013). They improve the capacity of the blood to carry oxygen and deliver it to the body's tissues. Blood transfusions are effective in preventing organ damage in patients with frequent, severe anemia (Pule & Wonkam, 2014). While transfusions are a valuable treatment option with life-saving results, they often lead to excessive iron in the blood. This iron can deposit in major organs causing serious damage that is often asymptomatic until the threat of organ failure is reached. Consequently, it is recommended that blood transfusions should be reserved for life-threatening complications (Kanter & Kruse-Jarres, 2013).

1.3.3 Nutritional Supplements

Folic acid is necessary for red blood cell production and growth. Due to the shortened lifespan of sickled red blood cells, SCD patients have a high red blood cell turnover rate that can deplete folate stores. Folic acid supplementation is recommended for SCD patients to ease the burden of anemia by promoting the growth of new blood cells (Ndefo et al., 2008). In addition, zinc is sometimes recommended in patients showing signs of zinc deficiency such as fatigue or multiple infections (Alli et al., 2014). While iron overload is often a problem for patients that receive transfusions regularly, those that are not transfused often may become iron deficit and need supplementation (Alli et al., 2014).

1.3.4 Preventive Strategies

In order to prevent potentially life-threatening infections, pediatric SCD patients are often prescribed a penicillin prophylaxis regimen. This is especially important for reducing mortality in children under five. All routine childhood vaccines are recommended along with additional immunization against *Streptococcus pneumoniae* and *Haemophilus influenzae* (Amid & Odame, 2014). In malaria endemic areas bed nets and indoor residual spraying are important in limiting a SCD patient's exposure to mosquitos (Ansong et al., 2013).

1.4 Variance in Severity

Although SCD is a monogenetic disease, patients sharing the same genotype display a substantial variation in phenotype that is not well understood. Several factors are thought to contribute to the range of symptoms experienced by patients. SCD patients in high-income countries typically have lower early mortality rates and less severe complications than those in many sub-Saharan African countries. Health system infrastructure, treatment capabilities, and access to medication are a huge part of this disparity (Tewari, Brousse, Piel, Menzel, & Rees, 2015). However, patients with access to the same medical facilities and treatment options also display wide varieties of phenotypic outcomes as well which could be attributed to genetic factors. One major pathophysiological factor in determining disease severity is a high HbS concentration (Steinberg, 2009). As a result, patients with HbSS rather than other variations of SCD, such as SC, have higher HbS concentrations and therefore usually display more severe clinical phenotypes. In contrast, high HbF levels are thought to reduce the severity of SCD by preventing the formation of HbS (Ndefo et al., 2008). HbF levels can be genetically linked to the specific haplotype associated with HbS (Bitoungui, Pule, Hanchard, Ngogang, & Wonkam, 2015). There are five main haplotypes described across the beta-globin gene cluster. The four associated with HbS in Africa are the Benin, Bantu/Central African Republic (CAR), Senegal, and Cameroon haplotypes. The India and/or the Arabian Peninsula (Arab/Hindu) haplotype is associated with HbS in parts of

Asia (Bitoungui et al., 2015). Studies have shown differences in health outcomes associated with specific haplotypes. For example, the Senegal haplotype has been associated with fewer hospitalizations and painful episodes whereas the Bantu haplotype has been associated with the highest incidence of organ damage and renal failure (Steinberg, 2009). In addition to health systems capabilities and genetic influence, several environmental factors have also been tied to disease severity such as temperature, air quality, wind speed, and altitude (Tewari et al., 2015). Environment also plays a role in determining a SCD patient's exposure to pathogens.

1.5 Psychosocial Stresses of Sickle Cell

The culmination of stress due to the chronicity and severity of disease symptoms, frequent interactions with health care systems, and fear and uncertainty about disease outcome can lead to substantial stress in both SCD patients and their family members. The inability to cope with this stress can negatively impact the quality of life of those affected (Mitchell et al., 2007). Social and emotional support from others has been shown to have protective benefits for health (Reblin & Uchino, 2008). For those coping with SCD, support from others has proven beneficial. In the US, studies suggest that improving SCD social support through family-centered interventions can benefit psychological adjustment and family relations (Gold, Treadwell, Weissman, & Vichinsky, 2008). In addition, children with SCD that receive higher levels of parental support have significantly decreased depressive symptoms and a better quality of life

(Sehlo & Kamfar, 2015). In low-resource settings, patients and caregivers are faced with enormous financial, interpersonal, and psychological burdens as they deal with SCD in areas with inadequate healthcare services; increased social support has been recommended to alleviate these burdens on SCD caregivers (Adegoke & Kuteyi, 2012). Improving the social and medical support that patients and caregivers receive would prove beneficial in coping with the psychosocial stressors of SCD.

1.6 Global Burden

SCD is the most common life-threatening monogenic disorder in the world. About 7% of the world's population carries the sickle cell trait (SCT) and 300,000–400,000 affected children are born each year (Aygun & Odame, 2012). In Africa, where about 85% of SCD cases are estimated to occur, 50–80% of infants born with HbSS die before the age of five years old (Grosse et al., 2011). The global distribution of sickle cell trait has been influenced by prevalence of *Plasmodium falciparum* malaria (F. B. Piel et al., 2010). People with HbAS have protection against severe forms of malaria, which influences the pattern of natural selection to favor sickle cell trait in areas where malaria is a major cause of death. As a result, SCD is most prevalent in areas with endemic malaria (Herbert Opi et al., 2014).

Cameroon is a lower-middle income country with a particularly high burden of SCD with 2-3% of its 22 million person population affected (A. K. Njamnshi et al., 2006). Despite the high prevalence of SCD, Cameroon lacks a specialized sickle cell center and

neonatal screening. While the country does have a national sickle cell control program, it is largely unimplemented leaving families to cope with high medical costs and inadequate treatment resources (Wonkam & Hurst, 2014).

In contrast, South Africa is an upper-middle income country with a naturally low prevalence of SCD. It has seen an increasing number of affected immigrants over the past decade (Wonkam et al., 2012). Cape Town in particular has a multitude of resources for sickle cell patients including a free of charge, specialized sickle cell clinic. Patients at this clinic have been shown to display less severe symptoms than sickle cell patients in other areas of sub-Saharan Africa indicating that a more developed social and medical resource network may lead to a more moderate phenotype (Wonkam et al., 2012).

By comparing the experience of those affected by SCD between these two countries, multiple variables can be examined. South Africa and Cameroon provide a comparison of vastly different healthcare systems dealing with different levels of disease prevalence. SCD is an illness with a standardized genotype among those affected and limited treatment options. Therefore, comparisons of different patient environments could prove useful; a SCD patient's environment will likely have a major effect on their health outcome. It is then crucial to understand the impacts of differences in available support, both medical and social. These differences could strongly influence the disease burden for those with SCD in these countries. By understanding differences in support

and the impact on health through a comparison of these two countries, the variance in SCD severity could be better understood overall.

1.7 Study Aims

This study focuses on understanding the burden of disease faced by SCD patients and their families in Cameroon and South Africa, and the role of social and medical support networks in their disease outcomes.

AIM 1: Determine the perceived burden of SCD experienced by sickle cell patients and their families in each location.

AIM 2: Identify the types of social and medical support received by sickle cell patients and their families in each location.

AIM 3: Assess relationships among social support, medical support, and disease burden within and between the study locations.

2. Methods

This mixed methods study used questionnaires and semi-structured interviews to quantitatively and qualitatively explore SCD burden and support systems in South Africa and Cameroon. The survey collected information on demographics, medical history, and financial factors. The interview focused on questions about the impact of SCD on daily life, the various sources of support, perceptions about SCD, and recommendations to ease the disease burden.

2.1 Setting

This study took place in three cities within two African countries: Cape Town, South Africa; Yaoundé, Cameroon; and Limbe, Cameroon. The interviews were conducted in four different hospitals: two hospitals in Cape Town, one in Yaoundé, and one in Limbe.

Groote Shuur Hospital (GSH) is a public hospital located in Western Cape offering tertiary and quaternary treatment. The hospital has multiple treatment capabilities including outpatient clinics, an Intensive Care Unit, and diagnostic and research laboratories. Patients at this hospital have been referred from a primary or secondary healthcare facility.

The Red Cross Memorial Children's Hospital (RCH) is a tertiary pediatric hospital in Cape Town that sees about 900 outpatients with complex hematological disorders each year. Due to an increasing prevalence of SCD, the Hematology/Oncology Unit has developed SCD-specific treatment guidelines. In 2009 the unit sponsored the development of satellite clinics at New Somerset Hospital and Victoria Hospital, which are two secondary-level hospitals in the Cape Town metropolitan area (Wonkam et al., 2012). Some participants interviewed at RCH have also received treatment at these satellite clinics. Patients that are relatively stable are often referred to these clinics whereas patients with complications are seen at RCH (Wonkam et al., 2012).

Yaoundé Central Hospital is a 381 bed tertiary level general teaching hospital with research and surveillance capabilities. The hospital staff includes 95 doctors and nearly 270 nurses and provides surgical care, obstetrics, gynecology, radiology, intensive care, and emergency services, as well as an outpatient clinic. This hospital serves a large population both within the city and across the country (WHO, 2009). While all interviews in Yaoundé were conducted in Yaoundé Central Hospital, some patients received care at SCD unit of the Mother and Child Centre of the Chantal Biya Foundation. This pediatric, tertiary health care facility is located next-door to Yaoundé Central. Its SCD unit was opened in December 2008 and deals with out-patient consultations, as well as the hospitalization of SCD children (Nansseu et al., 2015).

Limbe District Hospital is a small semi-urban hospital located in Southwest Cameroon. This 200-bed hospital has a 24-hour Emergency Department and a maternity unit with a 24 bed capacity. Two operating rooms handle cases requiring surgery (Tchounzou & Chichom-Mefire, 2015). The hospital lacks a blood bank and families are responsible for securing and paying for donors when a transfusion is necessary.

2.2 Participants

This study focused on SCD patients with HbSS; those with other forms of SCD were excluded. Eligible patients over the age of 18 completed a survey and interview on their own. If a patient with HbSS was under the age of 18, a caregiver completed the survey and interview on their behalf. Caregivers in this study were considered to be a

direct relative to the patient and lives with them and/or helps take care of them. Eligible caregivers were over the age of 18.

Participants were recruited and interviewed for this study from May to July 2015. The study was conducted in Cape Town during an 8 week period from the end of May to the beginning of July; the study was conducted in both sites in Cameroon in 2 weeks at the end of July. Recruitment was done largely through convenience sampling. In Cape Town the two hospitals used, Groote Schuur and RHC, have one designated SCD clinic day per week when all the SCD appointments are scheduled. On the clinic day usually between 0-5 patients would arrive for an appointment. Patients are seen on a rolling basis after arrival and often wait a few hours in the waiting room before they are seen. The patients and/or caregivers were approached during this waiting period and asked if they would like to participate in the study.

In Yaoundé, all interviews were conducted at Yaoundé Central Hospital where SCD patients are seen every day of the week. Patients and/or caregivers waiting for an appointment were approached to see if they would like to participate. In addition, some parents with admitted children and admitted patients that were feeling well enough to give an interview were asked to participate. Usually about 4-7 potential participants would be available for recruitment per day.

In Limbe, all interviews were conducted at Limbe District Hospital. Like the other sites, patients and caregivers waiting for their appointments were approached and

asked for their participation. Because a shorter amount of time was available to collect data at this site, 5 participants were also called over the phone by local health care providers and asked to come to the hospital specifically for the interview.

2.3 Procedures

After participant recruitment, surveys and interviews were conducted in a private room. In Cape Town an empty examination room was used at Groote Schuur and a consultation room was used at RHC. The principal investigator was the only interviewer present in the room for interviews in Cape Town. In Yaoundé a vacant doctor's office was used for interviews. The principal investigator was present for all interviews but due to language barriers, the interviews were conducted by a local doctor assisting with the study. Between two to four doctors were usually present in the room at the time of each interview. In Limbe the principal investigator conducted all interviews in an empty conference room at Limbe District Hospital. A local nurse assisted with a few interviews in which the participant and interviewer were having trouble communicating due to language comprehension issues.

All interviews were conducted the same way across the three sites. Subjects were approached by either the principal investigator in Cape Town and Limbe or a French-speaking research assistant in Yaoundé; all subjects that were approached to participate agreed to hear more information about the study. First, the study protocol and purpose were explained to the subject by reading from and summarizing the consent form. Each

participant was given a copy of the consent form detailing the process and expectations. One SCD patient in Cape Town did not wish to participate in the study and did not consent; the 57 individuals that consented to participate signed the consent form. In addition, participants could request more information about the results of this study, sickle cell disease, or information about new studies or support groups; they could indicate what they were interested in receiving directly on the consent form and write their contact information. There were 39 participants interested in some form of additional information. Those that did not request additional information did not leave any contact information.

After consent was received the two-part interview process consisting of the survey and interview began. The interviewer recorded the entire interaction. First, the interviewer read the questions from the paper survey to the participant and wrote their responses down. The survey portion typically lasted 10-20 minutes. Next, the interviewer indicated that the semi-structured interview would begin and that the participant could expand on their answers. The interviewer read questions from a paper interview guide to the participant. The interviewer did not take notes during this section and used the recorder to capture responses. The interviews did not adhere strictly to the interview guide and sometimes additional questions of interest or follow-up questions were asked. If a participant requested more information about a certain topic, they

received answers at the end of the interview. The semi-structured interviews typically lasted between 15-30 minutes.

In Cape Town and Limbe the recruitment, consent, and interview process was conducted in English. English was the first language of those native to South Africa and Limbe. For those that immigrated to South Africa, English was often a secondary language; while some immigrants had difficulty with a few questions due to language barriers, all participants considered themselves able to complete an interview in English. In Yaoundé, the entire process was conducted in French as that was their primary language. The English and French versions of the consent form can be found in **Appendix A**.

Participants in South Africa did not receive compensation. All participants were interviewed while they were waiting for an appointment, therefore no travel or productivity costs were incurred due to the interview.

Participants in Cameroon received 5000 CFA, or about $8.50, as compensation for their participation, because some of them did miss time from work and incur travel costs for the purpose of the interview. The funds were kept in a nurse's locked desk and patients went to her office after the interview to collect their compensation.

All study procedures were approved by the ethical review boards at Duke University and the University of Cape Town, Faculty of Health Sciences Ethics Committee.

2.4 Measures

This mixed-methods study involved a two-part interview process for each subject. Each interviewee, upon providing consent, was asked questions first from a structured survey, and then from a semi-structured interview guide. Questions in the interview and survey were developed using the results of SCD studies that focused on factors influencing the burden of disease for those affected by SCD such as hospital, financial, family, personal, and social factors; the results of studies identifying the psychosocial stressors of SCD on both patients and caregivers in Yaoundé were used in the development of questions that addressed relevant causes of stress previously identified in the research sites (Wonkam et al., 2014a, 2014b). Additionally questions were developed based on literature highlighting causes of stress, stigma, and health outcomes in SCD patients and their families in an African setting (Adegoke & Kuteyi, 2012; Alli et al., 2014; Dennis-Antwi, Culley, Hiles, & Dyson, 2011; Wonkam et al., 2012).

2.4.1 Survey

Two different survey formats were created: one for SCD patients giving the interview themselves and one for caregivers giving an interview about living with and/or taking care of a SCD patient. Both survey types were translated into French for interviews conducted in Yaoundé. Both survey formats contained four sections: demographic information, medical history, financial information, and hematological profiles. Surveys can be found in **Appendix A**.

2.4.2 Interview

Semi-structured interviews followed an interview guide to ensure that interviews were relatively standardized and comparable. Follow-up questions and additional questions were included at the researcher's discretion. This maintained the natural flow of conversation and allowed the freedom to inquire about topics specific to certain interviewees. The interview guide can be found in **Appendix A.**

2.5 Analysis

Each participant was assigned a code that corresponded to both their survey and semi-structured interview. Each code consists of three parts: two letters and a number. The first letter, either a "P" for patient or a "C" for caregiver, corresponds to the interview type. The next letter is either an "S" for South Africa, a "Y" for Yaoundé, Cameroon, or an "L" for Limbe, Cameroon. The numbers range from 1-57 and correspond to the overall interview number. The code "CS5" is therefore a caregiver in South Africa and the 5th overall interview whereas "PY40" is a patient in Yaoundé and the 40th overall interview.

The analysis process for the quantitative data involved first entering all paper surveys into an Excel spreadsheet. Each survey was entered individually and double-checked for errors. If a survey contained missing data or unclear answers, the corresponding interview recording was played to clear up any discrepancies. Codes were created to organize multiple choice answer choices into categorical data. Codes

were also created to summarize the extent of answers that were written-in such as "country of origin" and "occupation." No outliers were removed due to their representation of the sometimes extreme variation in SCD symptoms and experiences. Excel was used to analyze descriptive statistics for numeric data such mean, median, range, and standard deviation. Excel was also used to create tables displaying the survey data.

SAS was used for additional statistical analysis for a few survey questions of particular interest (Mushquash & O'Connor, 2006). The Excel sheet containing survey data was uploaded and items of interest were compared. Analysis was performed using the general linear model (GLM) procedure equivalent to a t-test, non-parametric test, and chi-square tests to test the selected factors for statistical significance.

Recordings of the semi-structured interviews were transcribed directly into Microsoft Word. While the entire interview process was recorded, in most cases the transcription started after the survey concluded and the semi-structured interview began. However, some responses of the survey portion were transcribed if the participant elaborated on their response beyond what could be recorded on the form. For example, when asked the survey question, "Are you currently employed?" if a participant answered with a story about being fired from a job due to SCD it was transcribed as well. However, if they gave a "yes/no" answer that could be indicated on the survey, their answer was not transcribed. All interviews conducted in French were

transcribed in French and then translated to English by a local bilingual doctor assisting with the research; he was familiar with both the medical terms and local dialect used to describe SCD in English and French.

After all interview transcriptions were complete, the Microsoft Word Document for each interview was uploaded into NVivo 11 as an internal source. First, nodes were created based on the questions asked during interviews as outlined by the interview guide. This created a uniform system to initially code the 57 interviews in a standardized way. Second, additional nodes were created that reflected some themes of interest that were not asked directly but that came up often. A codebook created in Excel was used to define the coding parameters for each node; this can be found in **Appendix A**. The initial coding process was completed using the line-by-line method; each interview was read individually and the answers to interview questions and general items of interest were assigned to their appropriate nodes. Next, text search queries were used to identify additional patterns found across interviews. Thematic analysis was used to organize the codes into categories and then into themes created by the researcher. This coding and analysis process was based on the strategies provided in *The Coding Manual for Qualitative Researchers* (Saldana, 2008). Representative quotes were selected that best encompassed the themes found as decided by the researcher. Quotes that were unique or that represented a different perspective were also selected.

Survey data and interview data were combined by importing an Excel spreadsheet of survey information into NVivo as a classification sheet. The sources were converted to case nodes and then classified by the demographic data found in the classification sheet. Then Matrix Coding Queries were conducted to identify relationships between interview responses and demographic data. These queries were also used to quantify certain themes and responses.

3. Results

A total of 29 patient interviews and 28 caregiver interviews were conducted which resulted in data for 57 SCD patients overall. There were 26 interviews in Cape Town (16 patient, 10 caregiver), 20 interviews in Yaoundé (10 patient, 10 caregiver), and 11 interviews in Limbe (3 patient, 8 caregiver). Since the interview participants were predominantly recruited through convenience sampling, the 57 patients were at the hospital for a variety of reasons. The majority, 32 patients (56%), were there for an appointment. There were also 9 for a pain crisis, 6 for general pain, 2 for an illness, 2 for a blood transfusion, and 1 for an unspecified reason. The breakdown of reasons that recruited participants were in the hospital is shown by research site in **Table 1.**

Table 1 Patients' Reason for Being in the Hospital

	Appointment	General Pain	Pain Crisis	Interview	Illness	Transfusion	Unknown	Total
Cape Town	22	1	0	0	2	1	0	26
Yaoundé	7	4	8	0	0	1	0	20
Limbe	3	1	1	5	0	0	1	11
Total	32	6	9	5	2	2	1	57

There were 17 interviews conducted at Groot Schuur; in addition to treatment at Groot Schuur 3 participants reported past treatment at RCH and 6 reported past treatment at other hospitals in South Africa. Of the 9 interviews conducted at RHC, only one participant reported past treatment at other hospitals in South Africa.

There were 20 interviews conducted at Yaoundé Central, with 19 of the patients receiving treatment there and 1 receiving treatment from only the Mother and Child

Centre. Two of the patients had received care at both Yaoundé Central and the Mother and Child Centre and 4 of the patients had received care at both Yaoundé Central and other Cameroonian hospitals.

Of the 11 interviews conducted at Limbe District Hospital, 9 participants reported receiving treatment at other hospitals in Cameroon.

3.1 Demographic Data

Of the 57 total patients, 28 (49%) were male and 29 (51%) were female. The 28 caregivers were mostly women with 18 (64%) females and 10 (36%) males. The breakdown of sex as well as age for the patients and caregivers is shown in **Table 2.** The caregivers were predominantly parents of SCD patients with 26 (93%) being the mother or father. One grandmother and one older sister were interviewed as caregivers as well. Most of the caregivers, 26 (93%), lived with the SCD patient; 21 (81%) of them reported attending the patient's appointments "always" or "very often" whereas 5 (19%;) reported attending "sometimes" or "rarely."

The age of SCD patients varied widely. The patients who provided their own data were an average age of 27.79 (±8.24) and ranged from 18-44 whereas the patients whose data were collected through caregiver interviews were an average age of 11.57 (±6.8) with a range of 1-33. The average overall patient age was 19.82 (±11.1) and ranged from 1-44 years old. The caregivers were an average age of 43.44 (±11.17) with a range of 18-67.

Table 2 Age & Sex of Patients & Caregivers

Patients				Caregivers	
	Patient Interviews	Caregiver Interviews	Total Patients		
Total	**29**	**28**	**57**	**Total**	**28**
Sex				**Sex**	
Male	10	18	28	Male	10
Female	19	10	29	Female	18
Age				**Age**	
average	27.79	11.57	19.82	average	43.44
SD	8.24	6.8	11.1	SD	11.17
median	24	10.5	18.5	median	42
range	18-44	1-33	1-44	range	18-67

The country of origin of participants in South Africa spanned 9 countries while all of the participants in Cameroon were also born in Cameroon. Of the 26 patients in South Africa, 12 (46%) were born in South Africa and 14 (54%) were born in other countries. Of adult patients in South Africa, 11 were immigrants which comprised the majority (69%; N=16). In contrast, 70% of the 10 pediatric SCD patients were born in South Africa, with 6 (86%) were born to immigrants. Of the caregivers interviewing on behalf of pediatric patients, 9 (90%; n=10) were immigrants and only 1 (10%; n=10) was born in South Africa. **Table 3** gives a breakdown of the countries of origin for South African patients and caregivers. The most prevalent country of origin for both patients and caregiver immigrants was the Democratic Republic of Congo (DRC) with 5 adult patients, 1 pediatric patient, and 5 caregivers being born there.

Table 3 Country of Origin

Country of origin for families coping with SCD in South Africa (n=36)				
	Patients			Caregivers
	Adult	Child	**Total**	**Total**
South Africa	5	7	**12**	**1**
DRC	5	1	**6**	**5**
Nigeria	1	0	**1**	**0**
Congo	1	0	**1**	**1**
Zambia	0	1	**1**	**1**
Zimbabwe	1	0	**1**	**0**
Angola	2	0	**2**	**1**
Lesotho	1	0	**1**	**0**
Tanzania	0	1	**1**	**1**
Total	16	10	**26**	**10**

The SCD patients that immigrated to South Africa have spent an average of 7.32 (±6.59) years in South Africa with a range of 1-25 years in the country. The caregivers have spent an average of 11.88 (±6.20) years in South Africa with a range of 6-25 years in the country.

Of the 57 participants, 52 (91%) identified as Christian; one participant further identified a Jehovah's Witness. There were 3 (5%) participants identifying as Muslim, one (2%) as non-religious, and 1 (2%) participant did not specify their beliefs.

Of the 29 patient interviewees, 21 (72%) had never been married. Four (14%) were currently married, one was divorced, one was widowed, one was engaged, and one did not specify their marital status. Six (38%; N=16) of the SCD patient interviewees in South Africa have either been married or engaged whereas in Cameroon only one patient interviewee (8%, n=13) had ever been married. Of the 28 total caregivers, 20

(71%) were currently married, 5 (18%) had never been married, 2 were widowed, and one was divorced.

With regard to the highest level of education, 14 (54%) of the SCD patient interviewees and caregivers in South Africa (n=26) reached the secondary education level, 3 (12%) reached college, and 5 (19%) reached university. Two participants reached a primary education level and 2 did not specify. Of the highest levels of education reached by participants in Cameroon (n=31), 4 (13%) had reached a primary education level, 21 (68%) reached secondary, and 6 (19%) reached university.

Of the patient interviewees, 12 (46%; n=26) were employed at the time of the study. Of the 14 (54%; n=26) that were not employed, 9 had never worked (6 of them were current students), and 5 had held a previous job but either stopped working or lost their job. Of the caregivers (n=27), 17 (63%) were employed. Of those who were not employed, 5 had held a previous job but either stopped working or lost their job, 2 were retired, 2 worked informally from their homes, and 1 was a student who had never worked.

Of the SCD patient interviewees, 9 (31%; n=29) of them had children. There were 6 patients with one child each, 2 patients with 2 children each, and one patient with 4 children. Of the SCD patients' 14 total children, none of them had SCD or had passed away for any reason. The caregivers' children will be analyzed in the context of SCD patients' siblings in a latter section.

Financial information for both countries was converted into US Dollars (USD); The South African Rand was converted at a 1 ZAR= 0.63 USD conversion rate and the Cameroonian Franc was converted at a 1 CFA= 0.0017 USD conversion rate. In Cape Town the average monthly income for SCD patients and their families (n=15) was $20 (±8.75). In Yaoundé (n=13) it was $121.68 (±69.48) and in Limbe (n=7) it was lower at $87.43 (±28.91).

Detailed data tables displaying the demographic survey results for each patient and caregiver can be found in **Appendix B.**

3.2 Medical History

3.2.1 Family History

Thirty-five (61.4%) of the 57 SCD patients reported at least one case of SCD in another family member. Of these 35 patients with a known family history of SCD, 22 (63%) were from Cameroon and 13 (37%) were from South Africa. Of the 13 South African patients with a family history of SCD, 8 (62%) were immigrants, 2 (15%) were children of recent immigrants, and 3 (23%) were adult patients native to South Africa. Of all the patients, 2 (4%; n=56) had a parent with SCD. Fifty patients (96%; n=52) had at least one living sibling. The SCD patients had a total of 174 siblings between them with an average of 3 (±2.18) siblings per patient. In South Africa patients averaged 2.6 (±1.8) siblings whereas in Cameroon patients averaged 4 (±2.3) siblings. Twenty patients (38%) reported having at least one sibling with SCD, for a total of 32 siblings with SCD. In

addition, 7 patients reported having a sibling that was suspected to have died from SCD; a total of 13 siblings were suspected to have died from SCD. Of the 187 reported siblings, those alive and those that had dead, 45 (24%) were thought to have SCD. In addition, 16 patients (28%) reported having either an aunt, uncle, or cousin with SCD, with a total of 21 relatives with SCD.

3.2.2 Age of Diagnosis

SCD patients in Cape Town had an average age of diagnosis of 7.95 (±7.17). In Cameroon the average age of diagnosis was 3.39 (±3.78). The ranked age of diagnosis was significantly lower in Cameroon (p =.0032). A breakdown of the age of diagnosis for patients across research sites can be found in **Table 4**.

Table 4 Age of Diagnosis

	Cape Town	Cameroon	Yaoundé	Limbe
	n=25	n=30	n=20	n=10
Average	7.95	3.39	3.24	3.7
SD	7.17	3.78	4.29	2.66
Range	0.33-26	0-16	0.25-16	0-9
Median	6	2	1.25	3

In South Africa, immigrants (n=13) had an average age of diagnosis of 9.31 (±7.25) whereas South African natives (n=12) had an average age of diagnosis of 6.48 (±7.09). There was an observable 2.83 year higher average age of diagnosis in patients born outside of South Africa as compared to native patients. However, the ranked age of diagnosis is not significantly higher (p=0.335) for immigrant patients in South Africa

when compared to patients born in South Africa. The breakdown of this comparison is demonstrated in **Table 5**.

Table 5 Age of Diagnosis in Immigrants Vs Native South Africans

	Born in South Africa	Born Outside South Africa
Age at Diagnosis	n= 12	n=13
Average	6.48	9.31
SD	7.09	7.25
Range	.33-24	2-26
Median	4	7

3.2.3 Frequency of Hospital Visits

The majority of SCD patients 79% (n=52) reported visiting the hospital at least once every 3 months. Of the 8 patients that reported visiting the hospital only once every 1-2 years, 7 (88%) were from Yaoundé. **Table 6** gives the breakdown of the frequency of hospital visits across the sites. **Figure 2** shows the frequency of hospital visits between the two countries.

Table 6 Frequency of Hospital Visits

	Cape Town	Yaoundé	Limbe	Total
	n=23	n=18	n=11	n=52
2+ per month	1	1	1	3
1x per month	6	4	7	17
1x per 2 months	6	0	0	6
1x per 3 months	8	5	2	15
1x per 6 months	1	1	1	3
1x per year	0	2	0	2
1x per 2 years	1	5	0	6

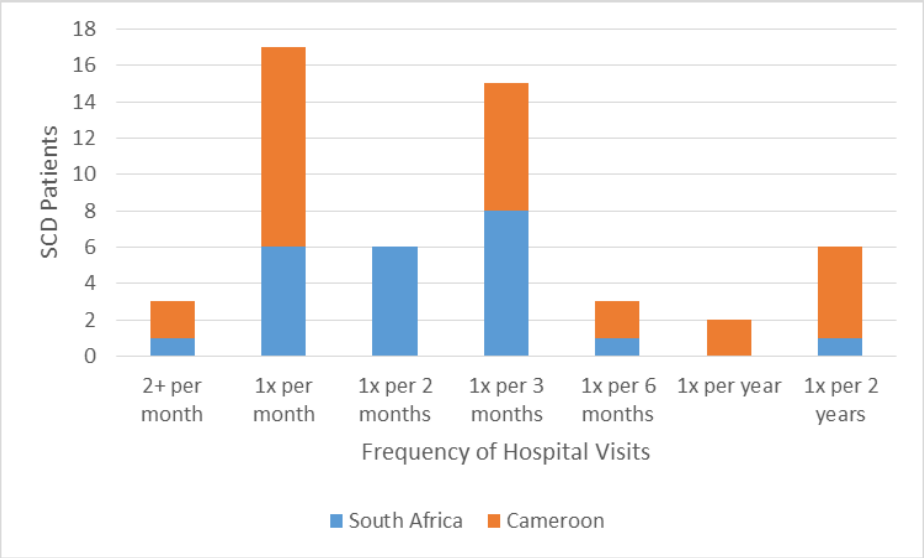

Figure 2 Frequency of Hospital Visits by Country

3.2.4 Hospital admissions per year

Patients in Cape Town (n=20) averaged 1.85 (±2.28) hospital admissions per year. In Cameroon (n=28) the overall average was 1.79 (±2.13). The number of hospital admissions per year was not significantly different between South Africa and Cameroon (p = .8728). On average, there were more hospital admissions in Limbe (n=10) with 3.4 (±2.76) per year, compared to Yaoundé (n=18) where there were 0.89 (±0.90) per year. The breakdown of annual hospital admissions across all the sites is provided in **Table 7**.

Table 7 Annual Hospital Admissions

	Cape Town	Cameroon	Yaoundé	Limbe
	n=20	n=28	n=18	n=10
Average	1.85	1.79	0.89	3.40
SD	2.28	2.13	0.90	2.76
Range	0-10	0-10	0-3	1-10
Median	1	1	1	2.5

3.2.5 Frequency of Pain Crises

Of the all the SCD patients, 13 (29%; n=45) experienced at least one pain crisis per month. Of these patients with the most frequent crises, 12 (92%; n=13) were from Cameroon. There were 11 patients (24%; n=45) that experienced a pain crisis every few months and 14 patients (31%; n=45) had a pain crisis a few times a year. There were 7 patients (16%; n=45) that experienced a pain crisis less than once a year and 6 of these patients (86%; n=7) were from South Africa. There were significantly more pain crises in Cameroon than in South Africa (p = .009). **Table 8** provides a breakdown of the frequency of pain crises across all sites and **Figure 3** demonstrates the frequency of crises between the two countries.

Table 8 Frequency of Pain Crises

	Cape Town	Yaoundé	Limbe	Total
	n=17	n=17	n=11	n=45
2+ per month	1	3	2	6
1x per month	0	1	6	7
1-2 per 3 months	2	9	0	11
1-2 per year	8	3	3	14
1x per 2 years	2	1	0	3
no crisis for 3+ years	3	0	0	3
never	1	0	0	1

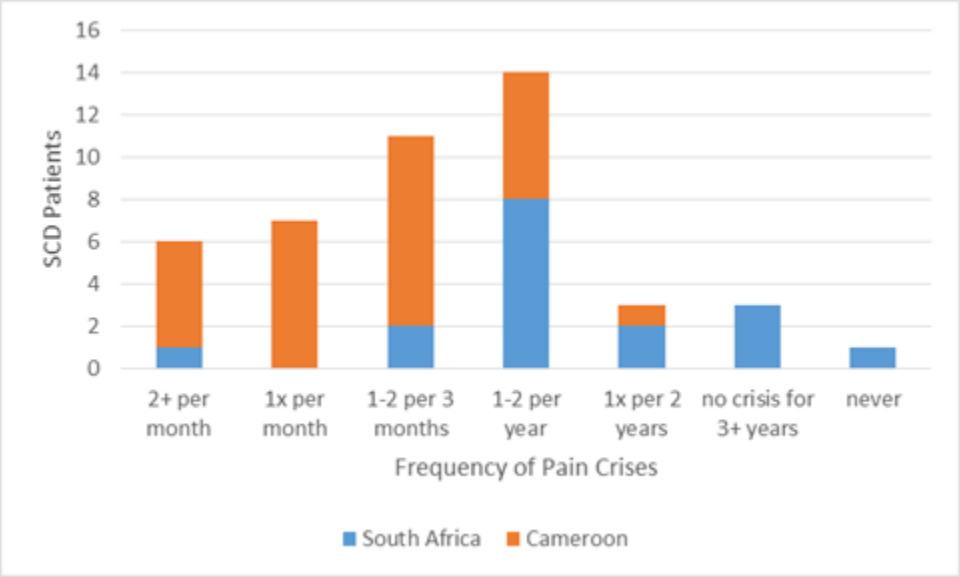

Figure 3 Frequency of Pain Crises by Country

3.2.6 History of Malaria

There were 38 SCD patients (75%; n=51) overall with a history of malaria. All 31 patients in Cameroon had contracted malaria in the past and had contracted it in Cameroon. In South Africa 7 patients (35%; n=20) had contracted Malaria in the past and all of them had contracted it in a different country. **Table 9** provides a breakdown of the history of Malaria in patients across both countries.

Table 9 History of Malaria by Country

	South Africa	Cameroon
Yes		
In current country	0	31
In a different country	7	0
No	13	0

3.2.7 History of SCD-Related Ailments

There was one SCD patient (2%; n=56) that reported having a stroke. Of the 57 total patients 20 (35%) had a history of infections, 4 (7%) had experienced problems with their spleen, 6 (11%) had a history of ulcers, 9 (16%) had experienced dactylitis, 5 (9%) had eye problems, 6 (11%) had gallbladder problems, 5 (9%) had experienced complications with treatment, and 6 (11%) reported arthritis. There were 25 (44%) patients that reported jaundice, with 22 (88%; n=25) of them being from Cameroon. When asked about additional health problems not mentioned in the survey, 3 patients reported hip necrosis and 2 patients reported gastritis.

3.2.8 Hydroxyurea Use

There were 16 patients (29%, n=55) currently taking hydroxyurea (HU) across all sites. Of these patients 15 (94%; n=16) were from South Africa. One patient in Cameroon (3%; n=30) was currently taking HU. Of the Cameroonian patients, 27 (90%; n=30) had never been offered HU and 2 (7%; n=30) chose not to take it because of infertility and health concerns. In Cape Town only 3 patients (12%; n=25) had never been offered HU. Of the South African patients not taking HU at the time of the study, 5 patients (50%; n=10) had been on HU previously but stopped taking it due to health concerns, side effects, and their prescription running out; two patients (20%; n=10) were too young to take HU. A complete breakdown of HU use across all sites can be found in **Table 10**.

Table 10 Hydroxyurea Use

	South Africa	Cameroon (Total)	Cameroon (by site) Yaoundé	Limbe	Patient Total
Yes	15	1	1	0	16
No	10	29	18	11	39
Reasons for Not Taking					
never offered	3	27	16	11	30
health reasons	1	1	1	0	2
side effects	3	0	0	0	3
too young	2	0	0	0	2
infertility concerns	0	1	1	0	1
prescription ran out	1	0	0	0	1

There were significantly less pain crises in patients that were using HU as compared to those that were not ($p = .0147$).

3.2.9 Additional Treatments Received

Of the SCD patients (n=57) across all sites 41 (72%) have received transfusions, 43 (75%) have received pain medications, 46 (81%) have received antibiotics, and 50 (88%) have received folic acid. There were 20 (35%) patients that had tried traditional medicine and all 20 of these patients were in Cameroon.

While reported separately in the survey, the treatments patients receive and SCD related illnesses they must avoid are interrelated. Patients and their caregivers must keep multiple treatments and prevention strategies in mind to maintain health. A few patients expanded on this connection by providing information outside the scope of the survey as illustrated in a longer response by a mother in Limbe:

> "Then they should always be on their folic daily. Yes, then on antibiotic also to prevent septicemia. Then too, you have to check them at least every month, because malaria like that cause them to have crisis. If they have malaria they will…fall into crisis and it will not be good. So…they should not be exposed to mosquitoes." (CL54)

In addition, patients also emphasized the importance of diet and hydration in maintaining their health and preventing pain crises. A patient in South Africa shared his strategy for staying healthy:

> "Just try and beat it, as in be two steps ahead. Before you feel you're going for an attack already have your vaccines, your tablets, your breakfast…Because for us it's very important to you know have constantly foods and energy and stuff with it, so I think if you're doing that you'll be fine really…In the morning I wake up, have my bottle of water, have a fruit, have a juice. Then later at work I have maybe a Weetbix or two, then lunchtime, throughout the day - just keep your body liquidized, not dehydrated—that's really bad. Because once you get dehydrated it can play a role - you start getting tired, dizzy." (PS14)

Detailed data tables displaying the survey results for patient medical history can be found in **Appendix B.**

3.3 Treatment Related Costs

The most common method of transportation to the hospital the day of the interview was by taxi with 32 (57%; n=56) families using that method. Taxis were particularly popular in Cameroon with 17 (89%; n=19) participants using taxis in Yaoundé and 9 (82%; n=11) in Limbe. In Cape Town (n=26) the means of transportation were fairly evenly distributed between bus (19%), the family's own car (12%), a friend or relative's car (19%), a taxi (23%), a train (15%), and walking (12%). Transportation methods to the hospital can be found in **Table 11**.

Table 11 Transportation to Hospital across Research Sites

	Bus	Own Car	Other Car	Taxi	Train	Walking
Cape Town	5	3	5	6	4	3
Yaoundé	1	1	0	17	0	0
Limbe	0	0	0	9	0	2
Total	6	4	5	32	4	5

Table 12 displays the monthly household income and the cost of roundtrip transportation for those using a bus, taxi, or train. In Cape Town the average transportation cost of $2.21 (±1.51) was 11% of the monthly household income. In Yaoundé, the average transportation cost of $3.53 (±3.17) was 3% of the monthly income. In Limbe the average transportation cost of $1.95 (±1.95) was 2% of the monthly income.

The cost of medical services was not relevant in South Africa where patients did not have to pay for care. **Table 13** compares monthly income with data collected on the cost of drugs, transfusions, and hospital bills in Cameroon**.** In Yaoundé the average monthly cost of medicine was $29.25 (±34.55), cost of transfusions was $32.22 (±12.62), and cost of hospital bills was $25.21 (±22.62). The cost of medicine, transfusions, and hospital bills consumed 24%, 26%, and 21% of the monthly income of $121.68 (±69.48) respectively. SCD medical care therefore comprises about 71% of the monthly income overall. In Limbe the average monthly cost of medicine was $42.50 (±20.82), cost of transfusions was $42.50 (±6.47), and cost of hospital bills was $21.28 (±10.71). The cost of

Table 12 Monthly Income & Transportation Cost (USD)

	Income	Trans-portation Cost
Cape Town	n=15	n=12
Avg	$ 20.00	$ 2.21
SD	$ 8.78	$ 1.51
Median	$ 18.90	$ 1.76
Yaoundé	n=13	n=19
Avg	$ 121.68	$ 3.53
SD	$ 69.48	$ 3.17
Median	$ 127.50	$ 1.70
Limbe	n=7	n=9
Avg	$ 87.43	$ 1.95
SD	$ 28.91	$ 1.95
Median	$ 85.00	$ 1.36

medicine, transfusions, and hospital bills took up 49%, 49%, and 24% of the monthly income of $121.68 (±69.48) respectively. SCD medical care then adds to about 122% of the monthly income overall.

Table 13 Cost of Medicine, Transfusions, & Hospital Bills in Cameroon

	Income	Cost of Medicine	Cost of Trans-fusions	Hospital Bills (yearly)	Hospital Bills (monthly)
Yaoundé	n=13	n=17	n=10	n=11	n=11
Avg	$ 121.68	$ 29.25	$ 32.22	$ 302.48	$ 25.21
SD	$ 69.48	$ 34.55	$ 12.62	$ 271.50	$ 22.63
Median	$ 127.50	$ 17.00	$ 29.75	$ 255.00	$ 21.25
Limbe	n=7	n=4	n=5	n=6	n=6
Avg	$ 87.43	$ 42.50	$ 42.50	$ 255.40	$ 21.28
SD	$ 28.91	$ 20.82	$ 6.47	$ 128.65	$ 10.72
Median	$ 85.00	$ 46.75	$ 42.50	$ 204.00	$ 17.00

Detailed data tables displaying the financial survey results for each patient's household can be found in **Appendix B.**

3.4 Hematology Profile

SCD-related lab values including red blood cell count (rbc), mean hemoglobin concentration (MCHC), white cell count (WBC), hemoglobin (Hb), hemoglobin A2 (HbA2), fetal hemoglobin (HbF), mean corpuscular volume (MCV), and platelets was recorded for each patient when available. A breakdown of test results by country can be found in **Table 14**.

Table 14 Hematology Profiles by Country

	WBCs ($10^3/mm^3$)	Hb (g/dl)	HbF (%)	MCHC (g/dl)	MCV (fl)	Platlets ($10^3/mm^3$)	RBCs ($10^6/mm^3$)	HbA2 (%)
South Africa	n=8	n=8	n=6	n=6	n=8	n=7	n=6	n=4
Avg	11.81	9.10	9.15	30.85	87.95	350.14	3.21	3.65
SD	4.53	1.51	7.92	4.92	10.60	100.33	1.08	0.76
Median	11.77	8.80	7.80	31.25	92.65	363.00	2.94	3.50
Range	4.31-17.1	7.1-11.8	1.2-22.9	22.1-36.5	67.2-96.8	199-511	2.24-5.3	2.9-4.7
Cameroon	n=16	n=17	n=1	n=14	n=16	n=16	n=15	n=0
Avg	15.25	6.89	17.60	32.60	88.16	354.44	2.56	-
SD	11.00	0.93	-	3.69	15.55	151.10	0.42	-
Median	13.05	7.20	17.60	32.80	82.50	379.50	2.72	-
Range	5.8-52.8	4.8-8.2	-	26-41.7	70-132	113-646	1.72-3.2	-
Total	n=24	n=25	n=7	n=20	n=24	n=23	n=21	n=4
Avg	14.11	7.60	10.36	32.10	88.09	353.13	2.75	3.65
SD	9.38	1.53	7.90	4.03	13.85	135.33	0.71	0.76
Median	12.50	7.30	10.90	32.30	86.75	379.00	2.74	3.50
Range	4.31-52.8	4.8-11.8	1.2-22.9	22.1-41.7	67.2-132	113-646	1.72-5.3	2.9-4.7

A detailed data table displaying the hematology profiles for each patient can be found in **Appendix B.**

3.5 Support Systems

The three main subjects covered in the interviews were social support, medical support, and burden of disease. **Table 15** describes common interview responses to the various aspects of support and disease burden perceived by interview participants; differences in common responses between the two countries should be noted.

Table 15 Common Topics and Themes Identified in Interviews by Country

	South Africa	Cameroon
Social Support		
Support of Close Family Members	Spouses aid caregivers; parents aid patients	Spouses aid caregivers; parents aid patients
Support of Other Relatives	Often update relatives; dislike sharing if relatives cause stress	Often update relatives; sometimes need to ask for assistance during crises
Friend Support	Some discuss with close friends for emotional support	Some depend on friends for assistance during crises
Friends' Understanding	Difficulty explaing to others; often avoid telling friends	Friends know of illness but may not understand it well; some feel pitied by friends
Support from other SCD Patients Outside of the Family	Very few participants knew others with SCD	All participants knew others with SCD; often speak casually with other patients; a few have attended support groups
Community Perception	Many do not know of the illness; people associate it with other blood-related illnesses	Many associate the illness with high severity and death
Myths & Misconception	All myths & misconceptions reported originated in other countries including: witch craft, disease ending at a certain age (usually 21), contagious, woman's fault	Caused by Ubange (witch craft), disease ending at a certain age (usually 21), contagious, woman's fault
Medical Support		
Health Care Providers	Very positive experiences	Mixed experiences
Cost of Treatment	Only indirect costs (transportation and food) were burdens	Very substantial source of stress; barrier to treatment
Perceived Effectiveness of Treatment	Satisfied with treatment overall	Many found treatment effective; many reported inefficient treatment as a major cause of stress
SCD Information Source	Received information from doctor and internet; many "Googled" SCD	Most only received information from doctors
Burden of Disease		
Daily Life with SCD	Pain, limitations in daily activities	Pain, limitations in daily activities
Hardest Part of SCD	Pain, uncertainity	Cost, pain, uncertainity, inadequate treatment
Sense of Control over Disease	Adult patients felt in control of the illness	Adult patients felt in control of the illness
Spititual Coping	Relied on God and spirituality for coping and relief from stress	Relied on God and spirituality for coping and relief from stress

3.5.1 Social Support

Participants' description of social support was defined by family support, friend & community support, and support from others with SCD outside their family. Differences in these themes between sites should be especially noted.

3.5.1.1 Family Support

Patients and caregivers in both countries reported receiving emotional support from family members. Patients in particular cited family members as sources of immense support such as this patient in Yaoundé: "They encourage me; everyone is always behind me, that's what still gives me strength" (PY29).

In Cameroon, financial support from family members was common and many parents relied on this support in times of crisis. One father in Cameroon described help from family members as essential to his son's survival:

> "If they were not there physically as [well as] morally, financially, the child..., we would have already lost him…The last crisis, I admit I had less than 30,000 francs as economy. But the bill at the end in ten days we didn't spent less than 300,000 francs. And we still continue to spend money, because it is something that will recur; so imagine what can happen in the head of a parent." (CY33)

Family members in Cameroon also aided in providing food and other means of support, especially during hospital admissions. Many stated that during admissions, "nobody is in the house to cook. So, food is coming in from the family members" (CL48). The family members would often to go the hospital with the patients; it is the family's responsibility to feed and provide basic care to admitted patients. According to

one patient, in addition to providing food "they usually administer the drugs to me, run around to look for nurses to give me some medicine" (PL47). Many patients also received help getting to the hospital during a crisis from those with cars.

Husbands and wives in both countries relied on each other over friends and family members. One father in Yaoundé said:

> "They do the best they can; those who have vehicles sometimes come and convey the child, I mean they organize to give a little boost. But that's not all the time, most is still based on me and my wife" (CY28).

In Cape Town where immigrant families did not have a large network of extended family nearby, spouses often relied on each other more heavily. One mother in Cape Town described making sure her husband could handle the child's illness in case she was not around:

> "Yes, I talk to my husband because you know the men, most of men, they don't take their shame too much. Because you see sometime the child, if he get sick, start to be sick and my husband sometime he panic and you just don't know what to do. And I told him, no you have to learn more because you don't know what is going to happen tomorrow for me. Because there's some time I'm not here you have to be strong. If the child is sick, don't panic. Just first take him and put him on your leg and find what is going wrong. If you know you can't manage you have to take him straight to the doctor." (CS10)

Patients with other family members with SCD sought support from them as well. Many cited that it was nice to talk to others with an understanding of what they were going through. One patient with 4 SCD siblings described the bond within her family:

> "It actually bring a lot of ... it put the family together firstly because you understand what the other person had because you have and when they are in pain you are there for them because you know that, you know, you won't be able

> to do certain things and it is really painful. So, you're there for them and you help them and sometimes it is so painful because, you know, it actually makes you cry and, you know, sometimes you can't blame God on all this. You just have to accept it" (PS26).

Many adult patients found that their SCD was less severe as compared to when they were children and as a result they required less outside support. They often described managing well on their own but still reaching out to their parents or other loved ones when necessary. One 40 year old patient in Yaoundé described this relationship with her relatives saying:

> "Currently as they know I'm already a big girl, well, they are not very concerned. They know that I manage that alone. When I need help they are there…they know I am often limited, so they come to my rescue. They financially often come to my rescue." (PY45)

Some patients described a lack of support from family members. Most of these patients felt that their family members "have their own problem, they have their own children" (CL55) and did not want to reach out and ask for assistance with their own problems. However, one patient in Cameroon felt that no relatives offered support due to stigma from the disease:

> "No one in my family supports me, because they do not consider me as a person. They are always saying that 'ha this one is not a person, she will die at any moment.' So it is useless to them. If I am hospitalized, only my mother will stay with me, with my sisters. So regarding family no one else can help me." (CL55).

3.5.1.2 Friend & Community Support

Both patients and caregivers in the two countries received various levels of support from friends and neighbors depending on how much they chose to disclose

their experience with SCD outside of their family. While many shared SCD news with close friends, many chose not to tell everyone. One mother in Cape Town described hiding it from friends to avoid the stress of their reactions to the illness:

> "Actually I never really want to like, to talk about it, because people they also scare me. When I want to know more they're like 'Oh getting older is going to be serious, it's dangerous.' Just don't want to talk more about it." (CS5)

One patient in Yaoundé described avoiding telling their friends due to fear of stigmatization:

> "I can't like talk about it to friends you know, because like if you know talk about it to friends you'll be marginalized; they won't understand that it's just like an inherited disease. Ya so you know, you might like have a lot of social conflict." (PY36)

In Cameroon, because the burden of care falls more heavily on the family, some families felt pressured to keep close friends in case they needed assistance, especially for transfusions:

> "Because they are always ready to come and donate blood. So you are forced to flatter, to always maintain good relationships with friends." (CY33)

Families also based their decision to reach out to friends on whether or not their friends would understand the illness. They could not simply say they had the disease without giving a full explanation of what it was. In South Africa, one patient lamented the lack of SCD understanding:

> "If I tell them, they will not understand what it is. So I have to explain it. Sometimes somebody asks me what it is. I have to explain it. Most of the people do not know actually what it is." (PS9)

One father in Cape Town described the difficulty in helping people to understand SCD. Because the illness is so uncommon many have never experienced it before and have nothing to relate it to, to aid in understanding. He stated:

> "People have absolutely no idea what sickle cell is. Even if I explain to them …they still wouldn't understand, I need to show them the physical book with graphics and pictures and all that kind of stuff in order for them to understand what Sickle Cell is. I think if people ask me about Sickle Cell, what is Sickle Cell, the easiest way for me to explain it to them is to say it is a blood disorder and you know then with the word blood disorder they connect that with leukemia, with blood cancer, with something like that. And then their first reaction will be 'ah shame' you know thinking that it is not manageable." (CS24)

In contrast, in Cameroon, the families found that many of their friends do "understand, since it is widespread, it is a disease that is not uncommon" (CY38). However, the knowledge was often limited to ideas like, "What people know is that it hurts too much, that they will not live long" (CY28). With only a basic understanding, many had misconceptions about what it truly meant.

There are several myths about SCD that have circulated within the communities that affect the level of support patients receive from those around them. One is that females with SCD cannot bear children and cannot make suitable wives. This idea was a common source of stigmatization that immigrants from the DRC experienced before immigrating to South Africa. Two immigrants described this stating that:

> "People used to say, she can't bear the children she will die, you can't marry her, as soon as you get a boyfriend, they going to go to the boyfriend and say you must leave her, she is going to get you in trouble, she is going to die." (PS26)

> "They always say, you know, you can't have kid, you won't live for long, you .... you know all bad things but ya but today I am so glad I have ... a doctor following up on me and I managed to ... to have two kids ... and I made it." (PS12)

When the genetics of SCD were not well understood, women were also sometimes solely blamed for the sick children in their family. One father in Yaoundé described the pressure he received from others to leave his wife because of this idea:

> "I said I couldn't leave my wife because she did not give birth to normal children. God gave me some normal ones and some of them sick; so I cannot reject my wife. You understand? Because we all are in the same situation." (CY32).

Some SCD patients also reported that others falsely believed that the disease is contagious:

> "They find it like a virus or something. Like you're gonna give it to me. Like you're sick, like sometimes you're fine and 'hey check that girl. She has sickle cell. She's anemic.'" (PS4)

In these instances, the patients themselves knew that their disease was not contagious but struggled when those in their community did not understand:

> "It's not contagious, most of the people they are afraid of, of getting sick as well. People they think that the disease is contagious and that we are normal like them." (PS13)

This idea can lead to stigma as people fear disease. Misconceptions about SCD being contagious also means that the true cause is not understood. For this reason one mother of a SCD patient did not know she could be a carrier and stated:

> "Because when my brother had it we didn't know that it genetic, we thought it was just a disease; that it just started." (CS25)

In Cameroon, many families found that their community believes SCD is caused by witch craft or sorcery referred to as *ubange*. This was reported by parents in both Cameroonian cities including this mother in Limbe:

> "I want other people to know that Sickle Cell is real because some people thought that was ubange because here in Africa people thought that these children are always sick and tiny like this, they think that it's ubange, that it is a witch." (CL49)

While community members believed that SCD was caused by witch craft, the families themselves knew that this was not the real cause as reported by this father in Yaoundé:

> "I speak, I always give advice, do not be discouraged, do not believe that this disease is a disease of witchcraft. This disease came because we did not do exams… Others say that sickle cell disease is sorcery. (CY43)

This belief that SCD is caused by witchcraft or a curse is damaging for a SCD patient's role in their community. They can be regarded as societal outcasts due to the stigma that they are cursed and will not live long. This is described by a father in Yaoundé:

> "Some people think it is witchcraft, others think it is a curse. That it is a disease that when it happens your life is meaningless. You are called to die, you are condemned to die, you're not considered in society, and you're a decreased person." (CY33)

3.5.1.3 Support from other SCD Patients Outside of the Family

In South Africa, the majority of participants did not know others with SCD outside of their family; only a couple knew other patients from the hospital or a few other immigrants in their community. When asked if they would like to know others

with SCD there were mixed responses. Some patients are more private and "just do not like to talk about it" (PS8). In general the older patients also did not see the benefit of meeting others because they were "used to it" (PS21). A 29 year old patient explained this position further stating that:

> "I can speak to anybody that's enquiring about it and wants to know what it is or what happens. But you know [I'm] at that age…when I get the symptoms I know what to do...so, I wouldn't really … want anybody to talk to." (PS14)

Because he can handle the clinical symptoms, this patient felt he did not need social support from others with the illness. However, caregivers and younger patients, namely those in their late teens and early twenties, did express interest in meeting others with SCD. Parents seemed interested as well "because this is all new" (CS24) to them and they wanted to know that their child's future might be like. Younger patients thought "it would actually be nice to hear other people's experiences" (PS15) and were interested in meeting those who personally understood their illness.

In contrast to the South African experience, nearly every participant in Cameroon knew of others with SCD, mostly from meeting them in the hospital. Many had friends and neighbors that were also affected. One father in Yaoundé knew several people with SCD and shared that, "in my village I know that are no less than 7 to 10, it's like an epidemic" (CY43). A few patients had attended a support group in Yaoundé, but none attended regularly. Despite irregular attendance one mother described her experience with the group:

> "It helped me because I saw those who were older, they even told us about those who were 70 years old, and it helped us. And they gave us tips how to keep, how to maintain a patient." (CY38)

Patients in Limbe also described attending occasional lectures and seminars where they were able to learn about SCD and discuss their experiences with other patients. One mother described these meetings:

> "Sometimes the doctor can call us here on the seminar and there we have a round table discussion people will be discussing their own way how the thing is happening to them, then I also hear and I will share my own with them, that's how we get our information" (CY49).

3.5.2 Medical Support

3.5.2.1 Health Care Providers

In Cameroon patients and families reported mixed reactions to the care provided by doctors and nurses which was summed up by the idea that, "There are always some bad apples" (CC28). Most felt respected overall but could still point to instances when a provider was "disrespectful, [and] welcomes us badly" (CY33).

In South Africa all participants felt respected by healthcare providers and had no problems. The only grievance was from an immigrant mother who felt the doctors did not fully understand SCD because it was less prevalent than her home country, the DRC:

> "When I got to South Africa I realized that the doctor in South Africa don't have much experience with it. I think back home there's more experience because the illness is more common than South Africa. Here it's like, they just have to read maybe for it. But back home, in the hospital, things like this they just know what, what, what. They know, knowledge." (CS5)

This mother also noticed a drop in health care provider knowledge and attention as her son transitioned from pediatric care to adult care and stated:

> "When I was at the Red Cross, that's where I learn a lot there about it. Because Red Cross, that's where he grew up. There was a lot of research, there was a lot of doctors that are concerned about it. More than [Groote Schuur]" (CS5).

However, despite the healthcare providers experiencing less SCD, all immigrants reported receiving better care in South Africa than in their home countries. An immigrant from Congo explained the faults in care in her country compared to what she was experiencing in South Africa:

> "In the Congo, people die quickly. The doctors are not so skilled or qualified and you have to pay and so many people are poor, so they are not accessed to the hospital. So, when you come, you just have to pay yourself. Some people they just get sick and they die. And the way they work is different from here, you see, so some people die quickly. And poor sanitation and the medicine is not so strong, or it is too few." (CS17)

3.5.2.2 Cost of Treatment

In Cameroon it was very difficult for families to afford the cost of treatment. Many indicated that all of their money went towards treatment leaving them unable to save. Sometimes they were unable to afford treatment when it is needed as stated by a grandmother in Yaoundé:

> "When there is no funding we cannot come to hospital. When we are asked for example to do exams, to pay particular medication, we must have means. And then, when he suffers it hurts and we cannot undergo that without being traumatized." (CY34)

The cost of all aspects of SCD are so expensive that some participants also described cost as a barrier to getting testing for SCD. A mother in Limbe stated:

> "[The testing] place is very expensive. The people don't even want to go and test so children are even there suffering; parents don't even know that it is sickle cell yeah just because the test is expensive." (CL49)

In addition to the already expensive task of raising a child with SCD, many families had multiple children with SCD as well which increased the burden as described by a patient in Limbe:

> "I know my parents will struggle to have the money to pay. It is a problem to pay the bills whenever we are in the hospital. When they struggle, they always do try to come up with the money to pay their bills in the hospital and to buy my drugs. Right now, my sister has been diagnosed also with the sickle cell, both of us. So, they are trying to pay the bill for two sicklers in the house." (PY39)

In Cape Town, many immigrants came from countries like Cameroon where the cost of treatment was very expensive. They noticed the differences in treatment capabilities and appreciated the accessibility of medication in South Africa:

> "You don't see that in Congo. You don't get medication every day like this. You get medication every day, every day. But in Congo we don't get that. It's expensive, we have to pay. And sometimes we don't have money to pay all the bills." (CS7).

3.5.2.3 Prognosis

Many of the patients and caregivers in Cameroon and those that immigrated to South Africa were told by both doctors and people in their communities about specific ages beyond which a child with SCD could not live. Many heard that they will not live past their early-twenties which was a major cause of stress as indicated by this mother in Yaoundé:

> "I was told they do not reach 21 years…You know that there are children on who you spend a lot of money. And when you put in your head that he will not reach 21 years, it's weird. It hurts a lot." (CY44)

Others heard that at age 21 SCD resolved itself. This is based on the belief that at this age the "disease will get finished" (CL51) or that they "cannot die then, the disease will not be as she was when she was young" (CL57).

Many immigrants realized that the 21 year old age limit they had heard in their home countries was not the expected prognosis in South Africa. This idea was expressed my several caregivers stating that:

> "Most of the time people say you can't have 21 years, he's gonna die before 21 years. But I come here to South Africa, I see a lot of anemic children, so different." (CS10)

> "If you go back to our countries the people who get this kind of sickness when they get to 18, 21 they are dying because of negligence you see." (CS7)

Despite their idea that, "People … don't mature there, they don't reach 25 years, in my country" when they "saw it in the adults" (CS7) in South Africa they realized the prognosis was different.

Immigrants to South Africa often expressed relief that they were dealing with SCD in Cape Town instead of their home country due to the differences in prognosis. One mother expressed:

> "In South Africa I can see you can manage. And I just say myself if this child was born there, in Congo, I don't know how I can cope, because it's not easy." (CS10)

3.5.2.4 Perceived Effectiveness of Treatment

Many patients and caregivers native to South Africa and Cameroon seemed satisfied with the efficiency of the treatment; however, they were often not able to compare it to anything else. Conversely, all immigrants to South Africa acknowledged that they received better overall care in South Africa compared to their home countries:

> "I am getting better treatment than in Congo. In Congo I can't afford to go to the hospital every time, because we have to go with money, if you don't have money they don't see you, but here at least they know that she is sickle cell and she need treatment and they give me medicine ya, for free." (PS12)

One patient that was a recent immigrant from Zimbabwe was prescribed HU after his first appointment at Groote Schuur. He was interviewed directly after his initial appointment and briefly interviewed again after his follow-up appointment two weeks later. When asked about his first two weeks on HU, he described a positive experience with his new treatment:

> "Ya, there are changes that I've noticed. And I haven't had any side effects with the medication that I'm taking. It's only for the first 15 minutes that I'm feeling like I want to sleep but after that, come back to be a normal person. Everything, can't complain. Ya, I'm seeing changes for sure. And I've noticed the other day it was very cold and I felt like my leg wanted to start again, the pain wanted to come back. As soon as I took those tablets the pain just disappeared. I've seen changes for sure." (PS1)

In Cameroon a few patients knew others outside the country which gave them a different perspective on the treatments available. They felt that they were not receiving equally effective medication:

> "The medication we used here is not efficient enough; because as I have friends who are abroad, when they give a treatment, foreign treatment is different from we have treatment here. If you are given a drug and then 30 minutes after you feel good, and the medicine you take here, you will have to wait maybe…45 minutes to one hour before feeling well, comfortable. So I find it ineffective." (PY45)

Many participants in Cameroon, despite not knowing about the existence of additional treatments still described the lack of effective SCD treatment as the most difficult part of the disease as stated by a mother in Yaoundé and father in Limbe:

> "The hardest thing is to know that there is not an appropriate treatment. It's really the most difficult thing to manage" (CY38)

> "The most difficult part is when you come to the hospital and the treatment is not responding." (CL48)

3.6 Burden of Disease

3.6.1 Daily Life with SCD

Participants in South Africa described pain as the hardest part of SCD. A significant burden described by caregivers in both sites was the impact of an unexpected pain crisis on the entire household. A father in Limbe describes the impact of his daughter's crises on daily activities:

> "We find the child, down in stress crying things first of all you feel depressed and the whole problem for the day because you are paying attention now to it. You cannot leave the house, the child is crying, so you have to think of what to do. Medications we try, if it does not work we have to take her to the hospital the whole day. If you want to do business [you cannot]." (CL48)

Similarly, a mother in Cape Town describes her frustrations when pain crises prevent the family from carrying out their plans for the day and the psychological toll crises have:

> "I can see the way she's getting sick every time, every time, every time. For me, it's not too good. It's not so good. I wish she could be like her sisters. She would be fine and I can be happy. But the way she gets sick, every time we must bring her. Sometimes she can get sick the time you are preparing to go to church. If that's the case the sisters must stay at home…because they must stay to look after her. Or myself I can stay to look after her and others can go to church. I'm not too much happy because of that." (CS2)

3.6.2 Sense of Control over Disease

Many patients, mainly those in South Africa, felt a sense of control over their illness through experience, resources, and education. They knew how to prevent a crisis and maintain their health. One 29 year old patient describes an almost casual approach to SCD as he's gotten older:

> "You know at that age whereby it doesn't really... bother me because I know when I get the symptoms I know what to do. It's not something that I would find that's life threatening you know because Sickle Cell is controllable. It's a disease where you can control the lifestyle you live you know. I don't smoke. Okay I do drink occasionally. When I feel I'm tired I do rest…when I feel weak I do contact [the doctor], come in, check for my blood, take it from there. So, ya it's something you can control." (PS14)

Participants in both South Africa and Cameroon felt a sense of control through God and an understanding that SCD was not necessarily terminal. One mother in Limbe expressed this, stating:

> "[God] is the one to take control. Whether you have Sickle Cell or not, you still die, some people are dying when they are any age and they don't have Sickle

> Cell…If you have Sickle Cell it does not mean that it is the end of their life. Sickle Cell is just like any ordinary disease, especially like this diabetes where people have to control." (CL57)

Patients that felt a sense of control over the disease reported feeling less overwhelmed than patients that felt it was unpredictable.

4. Discussion

The results suggest that differences in the strength of medical support systems influence the reliance on social support systems in a variety of ways. A strong medical support system provides patients with adequate treatment, education about their illness, respectful health personal, and financial costs that are within a family's ability to pay. When health care systems do not provide sufficient medical support, social networks of family and friends are used to complement the care and support necessary to treat SCD. A strong social support network provides patients and their families emotional support and understanding as well as assistance in receiving adequate treatment. Strong medical and social support systems seem to positively affect patient and caregiver experiences with SCD and their perceptions of disease burden.

Although South Africa does not have endemic SCD, its medical system is dealing with a rising number of SCD patients. In this study, 46% of the South African patients were born in-country. The proportion of SCD patients born in other countries (54%) is less than shown by an earlier study on the burden of SCD in Cape Town by Wonkam et al. that studied SCD patients at RCH between 2001-2010; this study showed that 93.1% of the patients were originally from other African countries, mainly the DRC (Wonkam et al., 2012). While the majority (69%) of South African adult patients in our current study were immigrants, only 30% of pediatric patients were born outside of South Africa. There appears to be a shift in the affected population's country of origin as

migration patterns lead immigrants from countries with endemic SCD to South Africa. Between 1960-2000, the number of migrants from countries with HbS allele frequencies higher than 10% increased from 3.1 million to 14.2 million; consequently, in this timeframe the estimated global number of migrants with HbS increased from about 1.6 million to 3.6 million (F. B. Piel et al., 2014). The results of this migration pattern changed the global distribution of SCD, and the countries receiving the majority of these immigrants, such as South Africa, are now seeing an increasing number of SCD patients born domestically. RCH has addressed this shift by strengthening its provided medical support to pediatric SCD patients through specialized treatment protocols; however, as the incidence of SCD births increases, the capacity for newborn screening and pediatric treatment will need to increase as well.

While 61.4% of SCD patients reported at least one case of SCD in another family member, 38.6% had no knowledge of relatives with SCD. Because only 4% of the patients had a parent with SCD, it was overwhelmingly transmitted by carriers who most likely had no knowledge of their trait status. In addition, 96% of the patients had at least one living sibling with 24% of all reported siblings suspected of having SCD as well. In environments with high SCD prevalence, a large family size and inadequate SCD education compound to exacerbate the problem. This is the case in Cameroon where many families reported multiple children with SCD. This points to faults in the educational components of the medical support system due to its inability to provide

accurate and timely diagnoses, family planning recommendations, or screening awareness in order to lower the incidence of SCD.

The burden of disease can be greatly influenced by the age of diagnosis because it signifies when SCD patients begin receiving SCD specific treatments and resources offered by the healthcare system. The age of diagnosis was significantly lower in Cameroon in comparison to South Africa, but still high across all sites. While the age of diagnosis for immigrants was not significantly higher when analyzed, there was an observable 2.8 year difference in age of diagnosis as compared to patients born domestically. This could suggest that South Africa may be diagnosing SCD earlier than surrounding countries, or that undiagnosed immigrants are falling through the cracks. Because the rate of childhood mortality for SCD is very high, especially in sub-Saharan Africa, it is essential to begin treatment as soon as possible after birth to improve survival rates. In areas with high ages of diagnosis, children with SCD often die before they are diagnosed (Tshilolo et al., 2008).

In Cape Town 60% of patients were on HU, with only 12% never being offered it as a treatment option. Of those in Cape Town that were not prescribed HU, 70% had a reason for not taking it including health concerns, side effects, or age. Although HU is the only available drug addressing the overall pathophysiology of SCD, it is not used for all SCD patients due to a variety of factors. In many settings the uncertainty of long term side effects can deter physicians from prescribing it (Segal et al., 2008). In Cameroon it

was particularly under-prescribed with 90% of patients never being offered HU as a treatment option. Unlike Cape Town, where 88% of patients were either taking HU or were not taking HU for a specific reason, those in Cameroon did not have the option. Additional barriers in Cameroon include access and availability. HU has not been officially recognized by the Ministry of Health in Cameroon as a treatment for SCD. This impacts the capacity for HU importation by local pharmacies. Cost is a significant factor as well; families already struggling to cover the cost of SCD treatment may not be able to afford an expensive, daily medication. In addition, the medical education of HU use is also not well established for health professionals in Cameroon. Laboratory infrastructure to test blood levels for patients on HU is also required which may present additional problems with availability and cost. The absence of HU as a treatment option substantially weakens Cameroon's medical support system.

While there was not a significant difference in hospital visits or hospital admissions between the sites, there were significantly more pain crises in Cameroon than in South Africa ($p = .009$). One likely explanation in the difference in pain crisis frequency is the use of HU; there were significantly less pain crises in patients that use HU as compared to those that did not ($p = .0147$). HU has been proven to reduce pain crisis frequency (Segal et al., 2008). Of those currently taking HU, 94% (n=16) were in Cape Town. Pain crises are an indicator of SCD severity and adult patients with more than 3 pain crises per year have been shown to have a decreased survival rate (Platt et

al., 1991). In Cape Town 18% (n=17) of patients experienced at least 3 pain crises a year whereas in Cameroon 75% (n=28) of patients reached this threshold. These factors indicate that the patients in Cameroon are experiencing more severe SCD symptoms as compared to patients in Cape Town. The absence of HU and resulting impact on disease severity highlights the relationship between a weak medical support system and an increased burden of disease.

The prevalence of malaria in Cameroon presented patients with an additional disease burden as compared to patients in South Africa. Malaria is considered a major cause of mortality for children in sub-Saharan Africa, especially those with SCD (Makani et al., 2010). Although children with SCD are less likely to contract malaria, they are at risk of a more severe clinical presentation when exposed to the parasite. The resulting parasitemia during hospitalization is associated with both severe anemia and death (Makani et al., 2010). Despite the decreased risk of SCD patients contracting malaria, the prevalence of malaria is so high in Cameroon that every SCD patient in this study had contracted it at least once. In contrast, anyone with a history of malaria in Cape Town was an immigrant who had contracted it in another country. This disease is another contributing factor to the higher overall burden of disease experienced by SCD patients in Cameroon compared to South Africa.

A previous study in Yaoundé that found adult SCD patients spent 40% of their monthly income on medicine and this financial burden was a significant cause of

psychological stress (Wonkam et al., 2014b). The stress caused by financial burden was also seen in this study as affected households in Yaoundé were shown to spend 71% of their monthly income on SCD treatment including medicine, transfusions, and hospital bills; in Limbe costs of treatment averaged 122% of monthly income. Participants in Limbe earned less money than those in Yaoundé, yet reported higher costs of medicine and blood. This could be possibly attributed to higher costs of accessibility in a rural location. The dependence on family and friends for financial support created an additional source of stress as described by participants in the interviews. Because those in Cameroon reported an inability to adequately save money, the stress of impending and unpredictable crises and their resulting economic strain was significant as well.

While the hematological data collected in Cape Town did not have a large sample size, the averages collected closely matched the hematological data collected during a previous study at RCH (Wonkam et al., 2012). The average HbF level of 9.15% is considered high for a SCD patient; this a good indicator of mild clinical severity and is likely attributed to HU use (Wonkam et al., 2012). The HbF level cannot be compared between sites as it is not regularly measured in Cameroon. In addition, the higher Hb and RBC levels in South Africa compared to Cameroon indicate less severe anemia and further indicate a less severe disease burden overall.

The main forms of social support were financial, emotional, and physical assistance during crisis. In this study caregivers reported relying heavily on spouses for

support whereas patients relied mostly on their parents. In South Africa, where financial burdens were significantly lower due to universal health care, social support was mostly emotional in nature. While patients that immigrated to South Africa had small support networks, none reported having to deal with SCD completely on their own and seemed to benefit from the support they were receiving from parents, close friends, or significant others. In Cameroon family support was especially important for health as it is the family's responsibility to feed and look after admitted patients in the hospital. Patients are also responsible for the full cost of their care and relied heavily on their friends and relatives for assistance, especially in cases of unexpected crises. In both sites, patients relied on family more at a younger age, and reported more independence as they matured. Older patients felt a sense of control over their illness and knew the steps to take to prevent crises as much as possible.

Community perceptions of SCD varied between sites. In South Africa, the low SCD prevalence meant that most of the SCD patients' friends and community members had never heard of SCD. In many cases they had nothing to relate it to, and patients struggled to help others understand their illness. In many cases, people with SCD chose not to disclose their illness to others as it is largely invisible in cases of mild phenotypes. Of those that reported experiencing community stigma and misconceptions, most were immigrants that had grown up in areas where SCD was very serious and patients experienced visible symptoms and early deaths. In Cameroon, most people knew of

SCD because it is very common. However, they often did not understand the cause or pathophysiology and instead understood it through myths including attributing causation to witchcraft and sorcery. Comparable stigmas in areas of high SCD prevalence have been reported in similar studies (Dennis-Antwi et al., 2011). Stigmas limit the social support available to patients and their families as they can lead to rejection from society.

Patients in South Africa were very satisfied with the medical services available to them. Immigrants were especially happy to receive treatment free of charge as many of them came from countries similar to Cameroon where they experienced a substantial financial burden. They also found the health care providers to be respectful overall. In Cameroon, patients were also satisfied with the treatment itself, but the financial burden often dominated their perception of care. The medical support was greater in South Africa as the medical system itself took care of many of the aspects that were left to family and friends in Cameroon such as treatment cost and basic care during hospital admissions. The addition of HU in the South African healthcare system also strengthened the medical support system by actively reducing pain crisis frequency leading to milder symptoms overall.

4.1 Implications for policy and practice

Early SCD detection is vital for improving disease outcomes, yet it is severely lacking in sub-Saharan Africa where SCD prevalence is highest. Newborn screening is

an essential tool not only for early SCD diagnosis, but for identifying those with sickle cell trait. Different newborn screening pilots have been tested in several countries including the DRC, Burkina Faso, Benin, Nigeria, and Ghana and have been shown to be cost-effective and useful in reducing patient morbidity and mortality (Tshilolo et al., 2008). Ghana has recently implemented a national new born screening protocol and it is essential that other sub-Saharan African countries follow suit. With the newborn screening programs, counselor training programs are recommended as well to ensure that adequate SCD education is provided alongside results (Treadwell, Anie, Grant, Ofori-Acquah, & Ohene-Frempong, 2014). A combination of national newborn screening programs and counseling upon receiving results would be highly beneficial in both South Africa and Cameroon.

Improved education for both patients and their communities is also essential. As seen in the myths and misconceptions that community members had about SCD, there is a deficit in community awareness about SCD. Awareness campaigns would not only reduce stigmatization surrounding the disease but could also help in prevention as many individuals did not understand the cause until experiencing SCD themselves.

Support groups may be beneficial in creating an environment of social support from others that understand the illness. Support groups would be especially beneficial in South Africa where patients had smaller support networks and little SCD understanding in their community. Most participants in South Africa reported knowing

no other SCD patients whereas nearly everyone in Cameroon knew of others with the disease. Support groups could supplement social networks with others that can relate to the illness. Support groups would also be an effective means of SCD health education where patients and caregivers could learn how to avoid crises and maintain good health. Although most patients in Cameroon did not attend the local support group, many reported attending a lecture or two hosted by the group and valued learning more about SCD.

4.2 Implications for further research

There is a paucity in SCD research that is especially evident in sub-Saharan Africa where SCD is the most prevalent. More research needs to be devoted to all areas of SCD (Grosse et al., 2011). From this study, it is apparent that more attention should be devoted to mitigating the rise of SCD in South Africa. Immigrants can easily fall through the gaps in healthcare systems that are unfamiliar with SCD and as a result can be diagnosed later in life and then treated with inadequate protocols. Without effective screening and education interventions, the frequency of HbS in South Africa will continue to increase based on global migration patterns (Frédéric B. Piel, Hay, Gupta, Weatherall, & Williams, 2013). Because of South Africa's strong medical support system, it could also serve as a model for other countries with no endemic SCD that are experiencing an increase SCD through global migration. It is therefore important to research the experience of immigrant families affected by SCD in South Africa to

determine what aspects of the healthcare system are beneficial in supporting life with SCD and what aspects can be improved upon.

In South Africa and Cameroon, programs addressing the psychological and socioeconomic burdens of SCD should be developed and tested, especially for patients without strong support networks. Because this chronic disease poses a huge burden for those without adequate support, programs addressing this issue could potentially improve health outcomes and quality of life. Country-specific programs will be essential to target the types of support necessary in each site. The cost of SCD treatment in Cameroon in particular needs to be addressed as it places an extremely high burden on families coping with this disease.

In addition to the need for medical support system strengthening through newborn screening in Cameroon and South Africa, the feasibility of introducing HU into resource-limited settings also needs to be examined. In Cameroon, this would be beneficial for patients suffering from excessive pain crises. Research would be required to assess providers' attitudes towards HU and the logistical challenges of its implementation. Studies demonstrating the effects of HU on patient health outcomes specific to Cameroon may also incentivize the government to issue an official treatment recommendation and consider providing financial assistance.

4.3 Study strengths and limitations

This study utilized a mixed-methods approach not previously used to study SCD in Cameroon or South Africa. It provides some of the first qualitative data addressing the SCD burden in both countries and assesses the experiences of both patients and caregivers. While SCD has been previously studied on a quantitative level in Yaoundé, this study provides some of the only data on SCD in Limbe (A.K. Njamnshi et al., 2006; Wonkam et al., 2014a, 2014b). Research on social support for SCD in an African setting is also very limited.

Furthermore, this study is one of the first to compare SCD experiences and disease burden between two countries. The different variables present in each location presented a unique opportunity to understand variations in SCD disease burden through the strength of support systems. This design would be useful in uncovering both positive and negatives aspects of approaches to SCD care in different locations as well. SCD could also be a good disease model for investigating differences in health systems across multiple sites. As a monogenic disease it is genetically straightforward with variation falling largely on the support and resources patients have access to; in addition, because of the limited treatment options the range of treatment variables would be limited. Because it is a chronic disease, patients' progress can also be followed over time. Therefore, not only can this multi-site SCD comparison model be beneficial

for improving SCD care globally, but it would also be beneficial for addressing general gaps in healthcare systems that could be highlighted through SCD treatment protocols.

Limitations include the design of convenience sampling as an imperfect approach for comparison between the sites. The patients in Cape Town were mostly in the clinic for appointments, while several patients in Cameroon were at the hospital for pain crises. Therefore, the populations selected for interviews could be skewed differently. In addition, some patients in Limbe were called in for an interview which did not fit the convenience sample method. However, because they were familiar with the hospital and often came in for appointments anyway, their recruitment was not likely to skew results differently from a patient there for a scheduled appointment. The problem of recall bias was evident as interviews were conducted. Medical records were only referred to for the collection of hematology results, therefore all other information came from the patients or caregivers directly. Questions about cost of treatment and frequencies of certain events were sometimes difficult for patients to recall.

Because the study did not sample across the entire population of the two countries, the ability to generalize results outside of the cities they were collected in is weakened. Although the participants interviewed do not completely represent their countries as whole, they did have a large variety of experiences and symptoms that provided a fairly comprehensive overview of SCD in those locations. Therefore, the data

collected, while not completely generalizable, will supplement the shortage of SCD research in both countries and possibly serve as a catalyst for further research efforts.

5. Conclusion

The burden of disease in Cameroon was higher than Cape Town based on both quantitative measures of severity and qualitative interviews. Compared to South Africa, Cameroon had less HU use which is the likely cause of the increased frequency of pain crises among Cameroonian SCD patients. The prevalence of malaria also created an additional health risk for those in Cameroon. Across all sites, SCD patients depended on family members for support. Those in South Africa relied more heavily on close family members whereas those in Cameroon relied on friends as well as family. The need for a larger social network in Cameroon was due to the high financial burden and the inability of many families to afford treatment without assistance. Because families in Cameroon are responsible for looking after their admitted family members with SCD, in addition to covering the full cost of treatment, support networks carried the added burden of providing food and assistance in the arrangement of medications and blood transfusions. These additional burdens are covered by a stronger overall medical support system in South Africa.

Due to the low natural prevalence of SCD in South Africa paired with the rising frequency of HbS trait through migration, newborn screening and education campaigns are essential in informing the largely unaware population about the cause and outcomes of SCD. Despite the high prevalence of SCD in Cameroon, patients and their families are

not receiving adequate support medically or financially and action must be taken to lessen the burden of disease and improve the prognosis.

Appendix A

Consent Form- English

Consent Form: Study on the Burden of Sickle Cell Disease
Code_____

This is a research study that will aim to understand the challenges faced by patients with sickle cell disease and their families. It is being conducted through Duke University (USA) and will look at participants in both Cape Town, South Africa and Yaoundé, Cameroon. We hope the resulting information about living with sickle cell disease in these two sites will improve sickle cell resources and make it easier to cope with this disease.

There will be a two part interview that will last approximately 30 minutes. The first part will consist of a questionnaire; the interviewer will read it to you and write down your responses. It will cover topics such as demographic information, medical history, and financial information. The second section will be an in-depth interview about life with sickle cell disease. The interviewer will use an interview guide to ask questions that you are encouraged to answer in detail. Your interview will be recorded.

The investigator may consult medical records to verify responses about health outcomes; only information relevant to the interview questions and sickle cell disease will be viewed.

There are minimal risks to participating in this study. Some interview questions may cause you discomfort or distress. Your participation is entirely voluntary and you may take a break, skip a question, or end the interview at any time. Participating in this study (or choosing not to) will not result in any changes to your current treatment or health services. You will not receive money or other incentives for participating in this study.

Your privacy is important to us. Your name will not be used in association with your interview answers or appear in any study reports. Any information you provide will be referenced using a code instead of your name or the name of the sickle cell patient. The paper questionnaire from Section 1 of the interview will be destroyed once the data is transferred to a computer. After the recording of your interview is transcribed, it will also be destroyed. All information collected from the interview (the questionnaire form from Section 1 and the transcribed recording from Section 2) will be stored on a secure online server.

If you have any questions or concerns, please feel free to ask them at any time. For further questions regarding this research you may contact Brittney Wittenbrink at britt.witt@duke.edu or Dr. Ambroise Wonkam at ambroise.wonkam@uct.ac.za. Questions regarding human subject research can be emailed to the Duke Office of Research Support ors-info@duke.edu. You will be given an additional copy of this form.

STATEMENT OF CONSENT:
If you agree to participate in this study, please sign the statement of consent below:

Printed Name of Participant_____

Printed Name of Sickle Cell Patient (if different than participant)_____

Signature of Participant_____ Date _____

Signature of Investigator _____ Date _____

Please indicate if you would like to be contacted in the future regarding any of the following:

- ☐ Study results
- ☐ More information on sickle cell disease
- ☐ Participation in future sickle cell studies
- ☐ New sickle cell disease education programs
- ☐ Support or outreach groups

- ☐ **No—do not contact me**

- Email Address_____
- Phone Number_____

Consent Form- French

Formulaire de consentement: Etude sur la charge de la drépanocytose
Code_____

Ceci est une recherche qui aura pour but de comprendre les défis auxquels font face les patients atteints de drépanocytose et leurs familles. Elle est menée par l'Université de Duke (Etats-Unis) et se penchera sur les participants à la fois à Cape Town, en Afrique du Sud et à Yaoundé, au Cameroun. Nous espérons que les informations recueillies à propos du fait d'avoir la drépanocytose dans ces deux sites permettront d'améliorer les ressources de la drépanocytose et permettre de faire face plus facilement à cette maladie.

Il y aura un entretien en deux parties qui durera environ 30 minutes. La première partie consistera en un questionnaire; l'interviewer vous le lira et écrira vos réponses. Il couvrira des sujets tels que les données démographiques, les antécédents médicaux, et les informations financières. La deuxième partie sera une entrevue en profondeur sur la vie avec la drépanocytose. L'interviewer utilisera un guide d'interview pour poser des questions auxquelles nous vous encourageons de répondre en détail. Votre interview sera enregistrée.

L'enquêteur pourrait consulter les dossiers médicaux pour vérifier les réponses en rapport avec votre état de santé; seules les informations pertinentes aux questions de l'entrevue et de la maladie seront considérées.

Il ya un risque minimal de participer à cette étude. Quelques questions d'interview peuvent provoquer un inconfort ou un chagrin. Votre participation est entièrement volontaire et vous pouvez prendre une pause, sauter une question, ou mettre fin à l'interview à tout moment. Participer à cette étude (ou choisir de ne pas le faire) n'entraînera pas de modifications de votre traitement ou de vos soins actuels. Vous recevrez 5000 FCFA comme frais de participation à cette étude.

Votre vie privée est importante pour nous. Votre nom ne sera pas associé à vos réponses d'entrevue et n'apparaitra dans aucun rapport d'étude. Toute information que vous fournirez sera référencée en utilisant un code à la place de votre nom ou du nom du patient drépanocytaire. Le questionnaire papier de la section 1 de l'entrevue sera détruit une fois les données transférées dans un ordinateur. Après que l'enregistrement de votre interview soit transcrit, il sera également détruit. Tous les renseignements recueillis lors de l'entrevue (la forme du questionnaire de la section 1 et l'enregistrement transcrit à partir de la section 2) seront stockés sur un serveur en ligne sécurisé.

Si vous avez des questions ou des préoccupations, s'il vous plaît sentir libre de les demander à tout moment. Pour plus de questions concernant cette recherche, vous pouvez contacter Brittney Wittenbrink à britt.witt@duke.edu ou Pr. Ambroise Wonkam à ambroise.wonkam@uct.ac.za. Les questions relatives à la recherche sur des sujets humains peuvent être envoyées par courriel au bureau de l'université de Duke de soutien à la recherche à ors-info@duke.edu. Vous recevrez une copie supplémentaire de ce formulaire.

Déclaration de consentement:
Si vous acceptez de participer à cette étude, s'il vous plaît signer la déclaration de consentement ci-dessous:
Nom du Participant_____

Nom du Patient (si différent de celui du participant) _____

Signature du participant _____ Date _____

Signature de l'investigateur _____ Date _____

S'il vous plaît indiquer si vous souhaitez être contacté à l'avenir en ce qui concerne les éléments suivants:

- ☐ Résultats de l'étude
- ☐ Plus d'informations sur la drépanocytose
- ☐ Participation à de futures études de la drépanocytose
- ☐ De nouveaux programmes d'enseignement de la drépanocytose
- ☐ Les groupes de soutien ou de sensibilisation
- ☐ **Non-ne pas me contacter**

  • Adresse Email_____
  • Numéro de Téléphone_____

Patient Survey

Interview Guide—Sickle Cell Disease Patient
Code_____

Section 1: Background Information

1. How old are you? _____
2. Gender?
   - Male
   - Female
3. What is your country of origin? _____
4. If not native- How long have you lived in this country?_____
5. Are you a new patient at this clinic?
   - Yes
   - No
6. At what age were you diagnosed with sickle cell disease?_____
7. Years since diagnosis_____
8. How many living children do you have? _____
   - How many of your children have SCD? _____
   - Have you lost any children due to SCD? How many?_____
9. How many siblings do you have? _____
   - How many have SCD?_____
   - Have you lost any siblings due to SCD? How many?_____
10. Do any of your family members have SCD? *Indicate how many
    - Mother
    - Father
    - Grandfather*_____
    - Grandmother*_____
    - Aunt*_____
    - Uncle*_____
    - Cousin*_____
    - Other_____
11. Are you married?
    - Yes
    - Never married
    - Divorced
    - Widowed
    - Other _____

12. What is the highest level of education you have completed?
    - □ No formal education
    - □ Primary
    - □ Secondary
    - □ University
    - □ Other_____
13. What is your occupation? _____
14. Are you currently employed?
    - □ Yes
    - □ No—held previous job
    - □ No—student/never worked
    - □ Other_____
15. What kind of area does the patient live in?
    - □ Urban
    - □ Rural
16. What is your religion?
    - □ Christian
    - □ Muslim
    - □ Non-religious
    - □ Other_____

Section 2: Health Information

17. How many times has the patient visited the hospital in the past month?
    - □ 2 or more per month
    - □ 1x per month
    - □ 1x every 3 months
    - □ 1x every 6 months
    - □ 1x every year
    - □ 1x every 2 years
18. Why did they visit the hospital today?
    - □ General pain
    - □ Pain crises
    - □ Appointment
    - □ Other_____
19. How many times were they admitted to the hospital in the past year?____
20. How often do you have pain crises?
    - □ 2 or more per month
    - □ 1 per month
    - □ 1-2 every 3 months

- 1-2 every year
- 1-2 every 3 years
- _____

21. Have they ever had a stroke?
    - Yes—How many?_____
    - No
22. Have they ever had Malaria?
    - Yes—In another country
    - Yes—In this country
    - No
23. What other health problems have they experienced due to sickle cell disease?
    - Infection
    - Splenic sequestration
    - Ulcers
    - Swelling in hands or feet
    - Delayed growth
    - Eye problems
    - Gallstone removal
    - Treatment complications (transfusion rejections, medication side effects etc.)
    - Jaundice
    - Arthritis
    - Other_____
24. Do they take Hydroxyurea?
    - Yes
    - No—but it was just prescribed this appointment
    - No—can't take it for health reasons
    - No—it was never offered
25. What treatments have they received in the past?
    - Blood transfusions
    - Pain killers
    - Antibiotics
    - Traditional medicine
    - Folic acid
    - Other_____
26. Where all do they receive treatment?

South Africa

- Groote Schuur
- Red Cross War Memorial Pediatric Hospital
- _____

Cameroon

- Yaoundé Central
- Mother & Child Center
- _____

Section 3: Financial Information

27. What is your average monthly household income?_____
28. How many people live in your household?_____
29. How many people in your household work? _____
30. How do you get them to the hospital?
    - Your car
    - Someone else's car—Who's?_____
    - Bus
    - Train
    - Taxi
    - Walking
    - Bike
    - Other_____
31. How much does transportation to the hospital cost?_____
32. Can you afford the sickle cell treatment recommended by the doctor?
    - Always
    - Very Often
    - Sometimes
    - Rarely
    - Never
33. What is the cost of drugs per month?_____
34. What is the cost of blood per month?_____
35. What is the yearly cost of hospital bills?_____

Section 4: Medical Records Information

36. WBC_____Date_____

37. Hemoglobin_____ Date_____

38. Fetal Hemoglobin_____ Date_____

39. MCHC_____ Date_____

40. MCV_____ Date_____

41. Platelets_____ Date_____

42. RBC_____ Date_____

Caregiver Survey

Interview Guide—Sickle Cell Disease Caregiver
Code_____

Section 1: Background Information
Interviewee:

1. How are you related to the sickle cell patient?
   - Father
   - Mother
   - Other_____
2. Does the patient live with you?
   - Yes
   - No
   - _____
3. How often do you bring the patient to their appointments?
   - Always
   - Very Often
   - Sometimes
   - Rarely
4. How old are you? _____
5. Gender?
   - Male
   - Female
6. What is your country of origin? _____
7. If not native- How long have you lived in this country?_____
8. Are you married?
   - Yes
   - Never married
   - Divorced
   - Widowed
   - Other _____
9. What is the highest level of education you have completed?
   - No formal education
   - Primary
   - Secondary
   - University
   - Other_____
10. What is your occupation? _____

11. Are you currently employed?
    - ☐ Yes
    - ☐ No—held previous job
    - ☐ No—student/never worked
    - ☐ Other_____

Patient

1. How old is the patient? _____
2. Gender of child with SCD?
    - ☐ Male
    - ☐ Female
3. What the patient's country of origin? _____
4. If not native- How long have they lived in this country?_____
5. Are they a new patient at this clinic?
    - ☐ Yes
    - ☐ No
6. At what age was the child diagnosed with sickle cell disease?_____
7. Years since diagnosis_____
8. How many siblings does the patient have? _____
    - How many of the children have SCD? _____
    - Have any children died due to SCD? How many?_____

9. Do any of the patient's family members have SCD? *Indicate how many
    - ☐ Mother
    - ☐ Father
    - ☐ Grandfather*_____
    - ☐ Grandmother*_____
    - ☐ Aunt*_____
    - ☐ Uncle*_____
    - ☐ Cousin*_____
    - ☐ Other_____
10. What kind of area does the patient live in?
    - ☐ Urban
    - ☐ Rural
11. What is your religion?
    - ☐ Christian
    - ☐ Muslim
    - ☐ Non-religious
    - ☐ Other_____

Section 2: Health Information

12. How many times has the patient visited the hospital in the past month?
    - 2 or more per month
    - 1x per month
    - 1x every 3 months
    - 1x every 6 months
    - 1x every year
    - 1x every 2 years
13. Why did they visit the hospital today?
    - General pain
    - Pain crises
    - Appointment
    - Other_____
14. How many times were they admitted to the hospital in the past year?____
15. How often do you have pain crises?
    - 2 or more per month
    - 1 per month
    - 1-2 every 3 months
    - 1-2 every year
    - 1-2 every 3 years
    - _____
16. Have they ever had a stroke?
    - Yes—How many?_____
    - No
17. Have they ever had Malaria?
    - Yes—In another country
    - Yes—In this country
    - No
18. What other health problems have they experienced due to sickle cell disease?
    - Infection
    - Splenic sequestration
    - Ulcers
    - Swelling in hands or feet
    - Delayed growth
    - Eye problems
    - Gallstone removal
    - Treatment complications (transfusion rejections, medication side effects etc.)
    - Jaundice
    - Arthritis
    - Other_____
19. Do they take Hydroxyurea?
    - Yes
    - No—but it was just prescribed this appointment
    - No—can't take it for health reasons
    - No—it was never offered

20. What treatments have they received in the past?
- Blood transfusions
- Pain killers
- Antibiotics
- Traditional medicine
- Folic acid
- Other_____

21. Where all do they receive treatment?

South Africa
- Groote Schuur
- Red Cross War Memorial Pediatric Hospital
- _____

Cameroon
- Yaoundé Central
- Mother & Child Center
- _____

Section 3: Financial Information

22. What is your average monthly household income?_____
23. How many people live in your household?_____
24. How many people in your household work? _____
25. How do you get them to the hospital?
- Your car
- Someone else's car—Who's?_____
- Bus
- Train
- Taxi
- Walking
- Bike
- Other_____

26. How much does transportation to the hospital cost?_____

27. Can you afford the sickle cell treatment recommended by the doctor?
- Always
- Very Often
- Sometimes
- Rarely
- Never

28. What is the cost of drugs per month?_____
29. What is the cost of blood per month?_____
30. What is the yearly cost of hospital bills?_____

Section 4: Medical Records Information

39. WBC_____Date_____

40. Hemoglobin_____ Date_____

41. Fetal Hemoglobin_____ Date_____

42. MCHC_____ Date_____

43. MCV_____ Date_____

44. Platelets_____ Date_____

45. RBC_____ Date_____

Interview Guide

Daily Life

- How does this disease impact your everyday life?
- What can't you do because of sickle cell disease?
- What is the hardest part of having sickle cell disease?

Family

- How does your family help you cope with sickle cell disease?
- Can you talk to your family about sickle cell disease when you need support? Do they understand?
- Do you talk to other family members with this disease?
    - How do you help each other?

Social/Friends

- Have you explained sickle cell disease to your friends? Do they understand what it is?
- How do your friends help you cope with sickle cell disease?
- Do you know others with sickle cell disease? How do you know them?
    - Do you talk to them about coping?
    - How do you support each other?
- What do people know about sickle cell disease in your community?
    - Are you embarrassed or ashamed because of this disease?
    - Does sickle cell disease affect how people view you?

Job/Studies

- How has sickle cell disease affected your job or school performance (loss of working hours or possibility of promotion)?
- Are employers/teachers understanding?

Health

- Are health care providers respectful? How do they treat you?
    - Do they listen to your concerns?
- Describe what happens during a pain crisis. What steps do you take?
- Is it hard to keep up with medication? Why?
- What would influence you to not seek treatment?
    - Tell me about the cost of treatment.
    - Do you find the treatment to be ineffective?
    - Do you feel that natural treatments are more effective?
    - Is the distance to the hospital an inconvenience?

Spiritual

- Do your religious/spiritual beliefs influence how you deal with sickle cell disease? If so, in what ways?
- How does your religious group support you?

General/Education

- Are there ever times when you are overwhelmed because of this disease? What do you do?
    - Do you ask others for help? Who do you ask?
    - What do you do for yourself?
- Do you feel that you've received adequate education about this disease?
    - From a doctor? Support group?
    - How do you get information about this disease?
    - What additional information would you like to receive?
- Is it different to have sickle cell in other countries? (Their country if they're an immigrant)
- What do you want others to know about this disease?
- Is there anything else you'd like to tell me about having sickle cell disease?

Codebook

daily impact	Daily struggles, everyday issues
limitations	What are you prevented from doing because of the disease? What can't you do?
hardest part	Hardest part of SCD, most difficult part
family help	How does family help? How does family support?
family talk	Do you talk to your family about SCD? What do you talk about?
family understand	Does you family understand what this illness is? Do they know what you're going through? Are they understanding about the difficulties?
family w/ SCD	Do you have family members with SCD
talk family w SCD	Do you talk to your family members with SCD about the illness? What do you talk about?
help family w SCD	How do your family members with SCD help you? How do you help eachother?
friends help	How do your friends help? Are your friends supportive?
friends understand	Do your friends understand what SCD is? Do they know what you're going through? Are they understanding about the difficulties?
friends talk	Do you talk to your friends about SCD? What do you talk about?
know others w SCD	Do you know others with SCD?
wants to know others w SCD	Would you want to know others with SCD? Why?
support group	Are you part of a support group? How does it help?
talk others w SCD	Do you talk to others with SCD? What do you talk about?
help others w SCD	How do others with SCD help you? Do you help them?

community SCD perspective	What do those in the community think about SCD? Do they understand what it is? Are they understanding of your struggles?
embarrassed ashamed	Are you embarrassed or ashamed because of the illness
job performance	How does SCD affect your job performance? Can you hold a job ok? Do you have to miss work a lot?
employer	Does your boss understand what SCD? Do they understand when you miss work?
school performance	How does SCD affect school performance? Are the grades ok? Are there problems with missing school?
teachers	Do teachers understand what it is? Are they understanding about missed class? Do they help the student catch up if they miss?
health care providers	Are the health care providers respectful? Do you ever have any problems with doctors or nurses? Are they competent in caring for SCD?
diagnosis	Anything related to the diagnosis, what initial information was given?
pain crisis	What happens during a pain crisis? Are there steps you take at home?
keep up with meds	How is it keeping with the medication schedule? Are there difficulties?
probs seeking treatment	Are there any problems getting treatment? Can you get the treatment you need? What would stop you from getting treatment
cost of treatment	What is the cost of treatment like? How does it affect your access
treatment effectiveness	Do you think the treatment you receive is effective? Does the medicine work?
traditional medicine	Have you ever tried traditional medicine or methods?

distance to hosp	Is the distance of the hospital from your house a barrier to treatment? Does the distance ever stop you from going to the hospital?
spiritual beliefs	How does your religion impact how you think about sickle cell? Do you pray about the illness?
help from church	How does your church help you? Does the church community support you?
overwhelmed	Are you ever overwhelmed by SCD? Are you very stressed or worried? What stresses you or worries you?
what do you do to cope	When you are stressed about the illness what do you do? Who do you talk to?
adequate SCD education	Do you feel you have enough education about SCD? What do you know about SCD?
SCD info source	How do you learn about SCD?
what info want to receive	What information would you like to know about SCD? What do you want the doctors to tell you more about?
SCD in other countries	Is it different to have SCD in other countries? Is it different in your birth country?
want others to know	What do you want others to know about SCD? Others in the community? Others with SCD?
SCD recommendations	What recommendations do you have for improving care for SCD patients and their families
general	Miscellaneous, interesting
misconceptions	Misconceptions or misinformation about SCD, either from interviewee or what others think about SCD
staying healthy	Things done to keep SCD patients healthy
testing	Did parents receive testing? Should parents receive testing?

household burden	How does SCD affect the entire household

Appendix B

Interview Code Descriptions

Interview Code	Interview Type	Interview Site	Interview Number	Interview Code	Interview Type	Interview Site	Interview Number
PS1	Patient	Cape Town	1	PY30	Patient	Yaoundé	30
CS2	Caregiver	Cape Town	2	CY31	Caregiver	Yaoundé	31
CS3	Caregiver	Cape Town	3	CY32	Caregiver	Yaoundé	32
PS4	Patient	Cape Town	4	CY33	Caregiver	Yaoundé	33
CS5	Caregiver	Cape Town	5	CY34	Caregiver	Yaoundé	34
PS6	Patient	Cape Town	6	CY35	Caregiver	Yaoundé	35
CS7	Caregiver	Cape Town	7	PY36	Patient	Yaoundé	36
PS8	Patient	Cape Town	8	CY37	Caregiver	Yaoundé	37
PS9	Patient	Cape Town	9	CY38	Caregiver	Yaoundé	38
CS10	Caregiver	Cape Town	10	PY39	Patient	Yaoundé	39
PS11	Patient	Cape Town	11	PY40	Patient	Yaoundé	40
PS12	Patient	Cape Town	12	PY41	Patient	Yaoundé	41
PS13	Patient	Cape Town	13	PY42	Patient	Yaoundé	42
PS14	Patient	Cape Town	14	CY43	Caregiver	Yaoundé	43
PS15	Patient	Cape Town	15	CY44	Caregiver	Yaoundé	44
CS16	Caregiver	Cape Town	16	PY45	Patient	Yaoundé	45
CS17	Caregiver	Cape Town	17	PY46	Patient	Yaoundé	46
CS18	Caregiver	Cape Town	18	PL47	Patient	Limbe	47
PS19	Patient	Cape Town	19	CL48	Caregiver	Limbe	48
PS20	Patient	Cape Town	20	CL49	Caregiver	Limbe	49
PS21	Patient	Cape Town	21	CL50	Caregiver	Limbe	50
PS22	Patient	Cape Town	22	CL51	Caregiver	Limbe	51
PS23	Patient	Cape Town	23	PL52	Patient	Limbe	52
CS24	Caregiver	Cape Town	24	CL53	Caregiver	Limbe	53
CS25	Caregiver	Cape Town	25	CL54	Caregiver	Limbe	54
PS26	Patient	Cape Town	26	CL55	Caregiver	Limbe	55
PY27	Patient	Yaoundé	27	PL56	Patient	Limbe	56
CY28	Caregiver	Yaoundé	28	CL57	Caregiver	Limbe	57
PY29	Patient	Yaoundé	29				

Demographic Data Profiles of Adult Patients

Inter-view Code	Sex	Age	Country of Origin	Education Level	Religion	Occupation	Employment Status	Marital Status	Number of Children
PS1	M	23	Zimbabwe	primary	-	food service	no- held previous job	never married	0
PS4	F	21	DRC	college	Christian	sales assistant	employed	engaged	0
PS6	F	23	Congo	secondary	Christian	-	no- never worked	never married	0
PS8	M	30	Nigeria	university	Christian	business	employed	never married	0
PS9	M	42	South Africa	secondary	Christian	business	-	married	4
PS11	M	26	South Africa	secondary	Muslim	-	employed	married	1
PS12	F	43	DRC	university	Christian (Jeho	teacher	no- held previous job	divorced	2
PS13	M	23	Angola	university	Christian	student	-	never married	0
PS14	M	29	Lethoso	secondary	Christian	insurance	employed	never married	0
PS15	F	24	South Africa	-	Muslim	pharm assistant	no- held previous job	married	0
PS19	M	18	South Africa	secondary	Christian	student	no- never worked	never married	0
PS20	M	18	Angola	secondary	non-religious	student	no- never worked	never married	0
PS21	F	28	South Africa	college	Christian	cashier	employed	never married	0
PS22	F	19	DRC	secondary	Muslim	cashier	employed	never married	0
PS23	M	44	DRC	college	Christian	-	no- held previous job	never married	0
PS26	F	31	DRC	secondary	Christian	sewing	employed	married	2
PY27	F	37	Cameroon	secondary	Christian	secretary	employed	never married	0
PY29	F	21	Cameroon	secondary	Christian	student	no- never worked	never married	0
PY30	F	43	Cameroon	secondary	Christian	housekeeper	employed	widowed	1
PY36	M	24	Cameroon	university	Christian	intern	no- never worked	never married	0
PY39	F	26	Cameroon	secondary	Christian	dressmaker	employed	never married	0
PY40	F	22	Cameroon	secondary	Christian	student	no- never worked	never married	0
PY41	F	24	Cameroon	secondary	Christian	trader	employed	never married	1
PY42	F	29	Cameroon	university	Christian	administrator	no- held previous job	never married	1
PY45	F	40	Cameroon	secondary	Christian	student	no- never worked	never married	1
PY46	F	18	Cameroon	secondary	Christian	-	-	-	0
PL47	F	24	Cameroon	university	Christian	student	no- never worked	never married	0
PL52	F	21	Cameroon	primary	Christian	hair dresser	training	never married	0
PL56	F	35	Cameroon	secondary	Christian	-	no- never worked	never married	1

Demographic Data Profiles of Caregivers & Pediatric Patients

	Patient Information				Caregiver Information									
Inter-view Code	Sex	Age	Country of Origin	Sex	Age	Country of Origin	Education Level	Religion	Occupation	Employment Status	Marital Status	Relationship to Patient	Lives with Patient	Frequency Attends Appts
CS2	F	6	South Africa	F	43	Congo	university	Christian	nurse	no- held previous job	married	mother	yes	always
CS3	M	9	South Africa	M	39	Angola	university	Christian	marketing director	employed	never married	father	yes	always
CS5	F	14	South Africa	F	43	DRC	-	Christian	fashion designer	no- held previous job	married	mother	yes	always
CS7	M	2	South Africa	F	-	DRC	secondary	Christian	-	employed	married	mother	yes	always
CS10	M	13	South Africa	F	42	DRC	secondary	Christian	cleaning service	-	married	mother	yes	always
CS16	F	4	South Africa	F	35	DRC	secondary	Christian	-	no- held previous job	married	mother	yes	always
CS17	F	13	DRC	F	18	DRC	secondary	Christian	student	no- never worked	never married	sister	yes	sometimes
CS18	M	10	Tanzania	M	46	Tanzania	primary	Christian	lifeguard	employed	married	father	yes	-
CS24	M	6	South Africa	M	43	South Africa	secondary	Christian	police detective	employed	living together	father	yes	always
CS25	M	10	Zambia	F	30	Zambia	secondary	Christian	-	no- held previous job	married	mother	yes	always
CY28	F	17	Cameroon	M	50	Cameroon	university	Christian	anthropologist	no- held previous job	married	father	yes	very often
CY31	M	16	Cameroon	M	62	Cameroon	secondary	Christian	joiner	employed	divorced	father	yes	sometimes
CY32	M	22	Cameroon	M	67	Cameroon	secondary	Christian	insurer	retired	married	father	yes	very often
CY33	M	11	Cameroon	M	31	Cameroon	secondary	Christian	business	employed	married	father	yes	very often
CY34	M	8	Cameroon	F	66	Cameroon	university	Christian	nurse	retired	never married	grandmother	yes	sometimes
CY35	F	13	Cameroon	F	40	Cameroon	secondary	Christian	trader	employed	widowed	mother	yes	rarely
CY37	M	8	Cameroon	F	32	Cameroon	university	Christian	teacher	employed	never married	mother	no	always
CY38	M	14	Cameroon	F	46	Cameroon	secondary	Christian	teacher	employed	married	mother	no	sometimes
CY43	M	15	Cameroon	M	56	Cameroon	primary	Christian	town hall worker	employed	married	father	yes	always
CY44	M	1	Cameroon	F	38	Cameroon	secondary	Christian	seller	-	married	mother	yes	-
CL48	F	24	Cameroon	M	57	Cameroon	secondary	Christian	banker	employed	married	father	yes	always
CL49	M	14	Cameroon	F	41	Cameroon	secondary	Christian	trainer	employed	married	mother	yes	always
CL50	M	9	Cameroon	M	43	Cameroon	primary	Christian	business	employed	married	father	yes	very often
CL51	M	7	Cameroon	F	42	Cameroon	secondary	Apostolic	chef	employed	married	mother	yes	always
CL53	F	33	Cameroon	F	50	Cameroon	secondary	Christian	accountant	employed	married	mother	yes	always
CL54	M	6	Cameroon	F	33	Cameroon	secondary	Christian	nurse	employed	married	mother	yes	always
CL55	F	7	Cameroon	F	42	Cameroon	primary	Christian	dry cleaner	employed	married	mother	yes	always
CL57	F	12	Cameroon	F	38	Cameroon	secondary	Christian	teacher	employed	widowed	mother	yes	always

Patients' Family History of SCD

Interview Code	Sex	Age	Number of Living Siblings	Number of Living Siblings with SCD	Number of Sibling Deaths due to SCD	Number of Parents with SCD	Number of Other Relatives with SCD
PS1	M	23	4	1	0	0	0
CS2	F	6	3	0	0	0	0
CS3	M	9	0	-	0	0	0
PS4	F	21	-	2	0	0	0
CS5	F	14	0	-	0	0	0
PS6	F	23	-	1	1	0	1
CS7	M	2	2	0	0	0	1
PS8	M	30	4	0	0	0	0
PS9	M	42	2	0	0	0	0
CS10	M	13	1	0	0	0	0
PS11	M	26	5	2	0	0	0
PS12	F	43	3	1	1	0	0
PS13	M	23	3	0	0	0	0
PS14	M	29	2	0	0	0	1
PS15	F	24	5	3	0	0	0
CS16	F	4	1	-	0	1	0
CS17	F	13	3	0	0	0	1
CS18	M	10	2	0	0	0	0
PS19	M	18	1	0	0	0	0
PS20	M	18	3	0	0	0	0
PS21	F	28	1	1	0	1	0
PS22	F	19	3	0	0	0	0
PS23	M	44	2	0	0	0	0
CS24	M	6	-	-	-	-	0
CS25	M	10	1	0	0	0	1
PS26	F	31	8	4	1	0	2
PY27	F	37	1	0	0	0	0
CY28	F	17	4	0	0	0	1
PY29	F	21	6	0	0	0	0
PY30	F	43	6	1	0	0	0
CY31	M	16	4	2	0	0	0
CY32	M	22	7	2	1	0	0
CY33	M	11	-	-	-	0	2
CY34	M	8	3	0	0	0	2
CY35	F	13	3	0	0	0	0
PY36	M	24	5	1	0	0	1
CY37	M	8	-	-	-	0	2
CY38	M	14	4	0	0	0	2
PY39	F	26	6	1	1	0	0
PY40	F	22	10	0	0	0	1
PY41	F	24	6	0	0	0	0
PY42	F	29	7	0	0	0	1
CY43	M	15	4	0	0	0	0
CY44	M	1	4	2	6	0	0
PY45	F	40	3	1	0	0	0
PY46	F	18	1	0	2	0	0
PL47	F	24	2	1	0	0	0
CL48	F	24	3	1	0	0	0
CL49	M	14	3	1	0	0	0
CL50	M	9	3	2	0	0	0
CL51	M	7	3	2	0	0	0
PL52	F	21	3	0	0	0	0
CL53	F	33	2	0	0	0	0
CL54	M	6	1	0	0	0	1
CL55	F	7	2	0	0	0	0
PL56	F	35	8	0	0	0	0
CL57	F	12	1	0	0	0	1

Patients' SCD Related Medical History

Interview Code	Sex	Age	Age at Diagnosis	History of Stroke	History of Malaria	History of Infection	History of Spleen Problems	History of Ulcers	History of Dactylitis	History of Eye Problems	History of Gall-bladder Problems	History of Treatment Compli-cations	History of Jaundice	History of Arthritis
PS1	M	23	5 years	X	yes	yes	X	yes	yes	yes	X	X	yes	yes
CS2	F	6	1 year	X	-	yes	X	X	yes	X	X	X	X	X
CS3	M	9	5 years	X	-	X	X	X	X	X	yes	X	X	X
PS4	F	21	4 years	X	-	X	X	X	X	X	X	X	X	X
CS5	F	14	2 years	X	-	X	X	X	yes	X	X	X	yes	X
PS6	F	23	8 years	X	-	X	X	yes	X	X	X	X	X	X
CS7	M	2	4 months	X	X	yes	X	X	X	X	X	X	X	X
PS8	M	30	7 years	X	yes	X	X	X	X	X	yes	X	X	X
PS9	M	42	13 years	X	X	X	X	X	X	X	X	X	X	X
CS10	M	13	5 months	X	X	yes	X	X	yes	X	X	X	X	X
PS11	M	26	10 years	X	X	yes	X	X	X	X	yes	X	X	X
PS12	F	43	2 years	yes	yes	X	X	X	X	X	X	X	X	X
PS13	M	23	20 years	X	X	X	X	X	X	X	X	X	X	X
PS14	M	29	2 years	X	yes	yes	X	X	X	X	X	X	yes	yes
PS15	F	24	5 years	X	X	yes	yes	X	X	X	yes	X	X	X
CS16	F	4	3 years	X	-	yes	X	X	X	X	X	X	X	X
CS17	F	13	8 years	X	X	X	X	X	X	X	X	X	X	X
CS18	M	10	7 years	X	X	X	X	X	yes	yes	X	X	X	X
PS19	M	18	12 years	X	X	X	X	X	X	X	X	X	X	X
PS20	M	18	17 years	X	X	X	X	X	X	X	X	X	X	X
PS21	F	28	24 years	X	X	X	X	X	X	X	X	X	X	X
PS22	F	19	6 years	X	X	yes	X	X	X	X	X	X	X	X
PS23	M	44	-	X	yes	X	X	X	X	X	X	X	X	X
CS24	M	6	2 years	X	X	X	X	X	X	yes	X	X	X	X
CS25	M	10	9 years	X	yes	X	X	X	X	X	X	X	X	yes
PS26	F	31	26 years	X	yes	X	X	X	X	X	yes	X	X	X
PY27	F	37	9 years	-	yes	X	X	X	X	X	X	yes	X	X
CY28	F	17	6 months	X	yes	X	X	X	X	X	yes	X	X	X
PY29	F	21	6 months	X	yes	X	X	X	X	X	X	X	yes	X
PY30	F	43	1.5 years	X	yes	yes	X	X	X	yes	X	yes	yes	X
CY31	M	16	2 years	X	yes	X	X	X	yes	X	X	X	yes	X
CY32	M	22	6 months	X	yes	X	X	X	X	X	X	yes	yes	X
CY33	M	11	7 years	X	yes	X	X	X	X	X	X	yes	yes	X
CY34	M	8	2 years	X	yes	yes	X	X	X	X	X	X	yes	X
CY35	F	13	11 years	X	yes	X	X	X	X	X	X	X	yes	yes
PY36	M	24	3 years	X	yes	X	X	yes	X	X	X	X	yes	yes
CY37	M	8	1 year	X	yes	yes	X	X	yes	X	X	X	yes	X
CY38	M	14	6 months	X	yes	yes	X	X	X	X	X	X	yes	X
PY39	F	26	3 months	X	yes	X	X	X	X	X	X	X	yes	X
PY40	F	22	2 years	X	yes	yes	X	X	X	X	X	X	yes	X
PY41	F	24	11 months	X	yes	X	X	X	yes	X	X	X	yes	X
PY42	F	29	8 months	X	yes	yes	X	X	X	X	X	X	yes	X
CY43	M	15	8 months	X	yes	X	X	X	X	X	X	X	yes	X
CY44	M	1	9 months	X	yes	X	X	X	yes	X	X	X	X	X
PY45	F	40	16 years	X	yes	yes	X	X	X	yes	X	X	yes	X
PY46	F	18	5 years	X	yes	yes	X	yes	X	X	X	yes	yes	X
PL47	F	24	1.5 years	X	yes	X	yes	X	X	X	X	X	X	X
CL48	F	24	1.5 years	X	yes	X	yes	X	X	X	X	X	X	X
CL49	M	14	9 years	X	yes	X	yes	X	X	X	X	X	X	X
CL50	M	9	3 years	X	yes	X	X	X	X	X	X	X	yes	X
CL51	M	7	6 years	X	yes	X	X	X	X	X	X	X	yes	X
PL52	F	21	-	X	yes	X	X	yes	X	X	X	X	X	X
CL53	F	33	-	X	yes	X	X	X	X	X	X	X	yes	yes
CL54	M	6	3 years	X	yes	yes	X	X	X	X	X	X	yes	X
CL55	F	7	3 years	X	yes	yes	X	X	X	X	X	X	yes	X
PL56	F	35	6 years	X	yes	yes	X	yes	X	X	X	X	X	X
CL57	F	12	4 years	X	yes	X	X	X	X	X	X	X	X	X

Patients' Medical Service Utilization

Inter-view Code	Sex	Age	Reason for Hospital Visit	Frequency of Hospital Visits	Frequency of Pain Crises	Annual Hosp. Admi-ssions	Current Use of HU	Use of Trans-fusions	Use of Pain Killers	Use of Anti-biotics	Use of Trad-itional Medicine	Current Use of Folic Acid
CS2	F	6	appointment	1x per 3 months	-	3	yes	yes	yes	yes	no	no
CS3	M	9	appointment	1x per 3 months	-	1	no	no	no	yes	no	yes
CS7	M	2	illness	1x per month	-	1	no	no	no	yes	no	yes
PS12	F	43	appointment	1x per 3 months	-	-	yes	no	yes	yes	no	yes
PS14	M	29	appointment	1x per 2 years	-	0	no	yes	yes	yes	no	yes
PS15	F	24	appointment	1x per month	-	-	no	yes	yes	yes	no	yes
CS16	F	4	illness	-	-	-	no	no	no	yes	no	yes
PS20	M	18	appointment	1x per 2 months	-	1	no	no	yes	no	no	no
CS25	M	10	appointment	1x per month	-	0	-	no	no	yes	no	no
PY27	F	37	general pain	1x per year	-	1	no	yes	yes	yes	yes	yes
CY32	M	22	pain crisis	1x per 2 years	-	0	no	yes	yes	yes	yes	yes
CY44	M	1	appointment	-	-	-	-	no	yes	yes	no	yes
PS11	M	26	appointment	1x per 3 months	1-2 per 3 months	3	yes	yes	yes	yes	no	yes
PS22	F	19	appointment	1x per 2 months	1-2 per 3 months	10	yes	yes	yes	no	no	yes
PY29	F	21	appointment	1x per month	1-2 per 3 months	1	no	yes	yes	yes	yes	yes
PY30	F	43	-	1x per 3 months	1-2 per 3 months	0	no	yes	yes	yes	yes	yes
PY36	M	24	appointment	1x per 3 months	1-2 per 3 months	1	no	yes	yes	yes	yes	yes
CY38	M	14	pain crisis	1x per 2 years	1-2 per 3 months	0	no	yes	yes	yes	no	yes
PY39	F	26	general pain	-	1-2 per 3 months	0	no	no	yes	yes	yes	no
PY40	F	22	general pain	1x per 3 months	1-2 per 3 months	-	no	yes	yes	yes	no	yes
PY42	F	29	general pain	1x per 3 months	1-2 per 3 months	2	no	yes	yes	yes	yes	yes
CY43	M	15	appointment	1x per 2 years	1-2 per 3 months	0	no	yes	yes	yes	no	yes
PY46	F	18	pain crisis	1x per 3 months	1-2 per 3 months	1	no	yes	no	no	yes	yes
PS1	M	23	general pain	-	1-2 per year	2	no	no	yes	yes	no	yes
PS6	F	23	appointment	-	1-2 per year	-	no	no	yes	no	no	yes
PS8	M	30	appointment	1x per 2 months	1-2 per year	1	yes	no	yes	yes	no	yes
CS10	M	13	transfusion	1x per month	1-2 per year	3	yes	yes	no	yes	no	no
PS13	M	23	appointment	1x per month	1-2 per year	1	no	yes	yes	yes	no	yes
PS23	M	44	appointment	2+ per month	1-2 per year	3	yes	yes	no	no	no	yes
CS24	M	6	appointment	1x per 3 months	1-2 per year	0	no	yes	no	yes	no	yes
PS26	F	31	appointment	1x per 3 months	1-2 per year	1	no	yes	yes	no	yes	yes
CY28	F	17	appointment	1x per month	1-2 per year	1	no	yes	yes	yes	yes	yes
CY31	M	16	pain crisis	1x per 2 years	1-2 per year	0	no	yes	yes	yes	yes	yes
PY45	F	40	pain crisis	1x per year	1-2 per year	1	yes	yes	yes	yes	yes	yes
CL49	M	14	interview	1x per 3 months	1-2 per year	1	no	yes	yes	yes	yes	yes
CL51	M	7	appointment	1x per 6 months	1-2 per year	2	no	yes	yes	yes	no	yes
PL52	F	21	interview	1x per month	1-2 per year	1	no	yes	no	no	no	yes
CS5	F	14	appointment	1x per 3 months	1x per 2 years	2	yes	yes	no	no	no	yes
CS17	F	13	appointment	1x per month	1x per 2 years	1	yes	yes	no	no	no	yes
CY34	M	8	pain crisis	1x per 2 years	1x per 2 years	0	no	no	yes	yes	no	yes
CY37	M	8	pain crisis	1x per 6 months	1x per month	3	no	yes	yes	yes	yes	yes
PL47	F	24	general pain	1x per 3 months	1x per month	-	no	yes	yes	yes	yes	yes
CL50	M	9	appointment	2+ per month	1x per month	3	no	yes	yes	yes	yes	yes
CL54	M	6	interview	1x per month	1x per month	4	no	no	yes	yes	no	yes
CL55	F	7	interview	1x per month	1x per month	3	no	yes	yes	yes	no	no
PL56	F	35	appointment	1x per month	1x per month	6	no	yes	yes	yes	yes	yes
CL57	F	12	interview	1x per month	1x per month	2	no	no	yes	yes	yes	yes
PS21	F	28	appointment	1x per 2 months	2+ per month	4	yes	yes	yes	yes	no	yes
CY33	M	11	appointment	2+ per month	2+ per month	2	no	yes	yes	yes	no	yes
CY35	F	13	appointment	1x per month	2+ per month	1	no	yes	yes	yes	no	yes
PY41	F	24	pain crisis	1x per month	2+ per month	2	no	yes	yes	yes	yes	yes
CL48	F	24	pain crisis	1x per month	2+ per month	2	no	yes	yes	yes	yes	yes
CL53	F	33	transfusion	1x per month	2+ per month	10	no	yes	yes	yes	no	yes
PS19	M	18	appointment	1x per 2 months	never	0	yes	no	no	no	no	no
PS4	F	21	appointment	1x per 6 months	no crisis for 3+ years	0	yes	no	no	no	no	yes
PS9	M	42	appointment	1x per 3 months	no crisis for 3+ years	-	yes	yes	yes	yes	no	yes
CS18	M	10	appointment	1x per 2 months	no crisis for 3+ years	-	yes	yes	no	yes	no	yes

Financial Profiles for Families Affected by SCD

Inter-view Code	Inter-view ode	Number of People Living in the Home	Number of People Working in the Home	Monthly Household Income	Transport to Hospital	Cost of Transport Roundtrip to Hospital ($)	Cost of Drugs per Month ($)	Cost of Blood per Month ($)	Annual Hospital Bills ($)	Ability to Afford Care
PS1	1	1	0	-	taxi	-	-	-	-	very often
CS2	2	6	2	-	train	$ 1.26	-	-	-	always
CS3	3	2	1	$ 31.50	own car	-	-	-	-	always
PS4	4	5	1	$ 28.35	other car	-	-	-	-	always
CS5	5	-	1	-	own car	-	-	-	-	always
PS6	6	8	1	-	other car	-	-	-	-	always
CS7	7	5	2	$ 18.90	taxi	$ 2.84	-	-	-	always
PS8	8	1	1	$ 31.50	other car	-	-	-	-	sometimes
PS9	9	9	1	-	taxi	$ 0.63	-	-	-	always
CS10	10	6	1	$ 10.40	bus	$ 3.15	-	-	-	always
PS11	11	3	-	$ 18.90	other car	-	-	-	-	-
PS12	12	-	-	-	bus	$ 1.64	-	-	$ 18.90	sometimes
PS13	13	2	-	-	train	$ 2.21	-	-	-	very often
PS14	14	4	2	$ 28.35	bus	$ 0.57	-	-	-	-
PS15	15	2	-	$ 25.20	other car	-	-	-	-	-
CS16	16	6	-	$ 12.60	taxi	$ 1.76	-	-	-	-
CS17	17	-	-	-	train	$ 1.76	-	-	-	-
CS18	18	9	-	$ 2.39	bus	$ 2.65	-	-	$ 4.41	always
PS19	19	4	1	-	train	-	-	-	-	always
PS20	20	4	2	-	walking	-	-	-	-	very often
PS21	21	6	4	$ 15.75	bus	-	-	-	-	always
PS22	22	6	1	-	walking	-	-	-	-	always
PS23	23	-	-	$ 10.08	walking	-	-	-	-	-
CS24	24	3	2	$ 15.75	own car	-	-	-	-	always
CS25	25	4	1	$ 25.20	taxi	$ 1.76	-	-	-	-
PS26	26	4	1	$ 25.20	taxi	$ 6.30	-	-	-	always
PY27	27	4	1	$ 212.50	taxi	$ 1.02	-	-	$ 255.00	-
CY28	28	6	2	$ 127.50	taxi	$ 8.50	$ 17.00	$ 34.00	$ 204.00	always
PY29	29	6	2	-	taxi	$ 1.02	$ 5.10	-	$ 510.00	always
PY30	30	3	1	$ 51.00	taxi	$ 1.19	$ 17.00	$ 51.00	$ 255.00	rarely
CY31	31	6	1	-	taxi	$ 1.70	$ 8.50	$ 30.60	$ 102.00	sometimes
CY32	32	15	0	$ 136.00	taxi	$ 8.50	$ 4.25	$ 42.50	$ 51.00	very often
CY33	33	6	2	$ 42.50	taxi	$ 3.06	$ 17.00	$ 51.00	$ 204.00	very often
CY34	34	5	1	$ 136.00	taxi	$ 2.04	$ 25.50	-	$ 306.00	very often
CY35	35	5	1	-	bus	$ 6.80	$ 6.80	-	-	always
PY36	36	4	3	$ 102.00	taxi	$ 1.19	$ 10.20	-	-	sometimes
CY37	37	4	2	$ 238.00	taxi	$ 2.04	$ 6.80	$ 20.40	-	always
CY38	38	7	2	$ 221.00	taxi	$ 1.70	$ 59.50	$ 28.90	$ 79.73	always
PY39	39	19	3	$ 51.00	taxi	$ 1.70	$ 59.50	-	-	-
PY40	40	7	2	-	-	$ 10.20	-	$ 14.45	-	always
PY41	41	6	2	$ 29.75	taxi	$ 8.50	$ 127.50	$ 20.40	-	very often
PY42	42	5	2	-	taxi	$ 1.70	$ 85.00	-	$ 1,020.60	always
CY43	43	9	1	$ 98.60	taxi	$ 3.40	$ 34.00	$ 28.90	$ 340.00	always
CY44	44	5	1	-	taxi	$ 1.70	$ 5.10	-	-	always
PY45	45	3	1	$ 136.00	taxi	$ 1.02	$ 8.50	-	-	sometimes
PY46	46	4	1	-	own car	-	-	-	-	always
PL47	47	6	2	-	taxi	$ 0.85	-	-	-	always
CL48	48	6	2	-	taxi	$ 6.80	-	$ 42.50	-	-
CL49	49	5	1	-	taxi	$ 1.36	-	$ 45.90	$ 204.00	sometimes
CL50	50	8	1	$ 85.00	taxi	$ 2.04	-	-	$ 204.00	sometimes
CL51	51	8	2	$ 127.50	taxi	$ 2.38	$ 34.00	$ 34.00	$ 204.00	sometimes
PL52	52	4	4	-	walking	-	$ 59.50	-	-	sometimes
CL53	53	7	2	$ 119.00	taxi	$ 2.04	-	$ 51.00	-	sometimes
CL54	54	5	2	$ 68.00	taxi	$ 1.02	$ 59.50	-	$ 255.00	sometimes
CL55	55	5	1	$ 42.50	taxi	$ 0.51	$ 17.00	-	$ 155.38	sometimes
PL56	56	9	1	$ 85.00	taxi	$ 0.51	-	$ 39.10	$ 510.00	rarely
CL57	57	4	1	$ 85.00	walking	-	-	-	-	rarely

Patient Blood Work

Interview Code	WBCs ($10^3/mm^3$)	Hb (g/dl)	HbF (%)	MCHC (g/dl)	MCV (fl)	Platlets ($10^3/mm^3$)	RBCs ($10^6/mm^3$)	HbA2 (%)
CS7	15.8	7.9	11.7	-	77	199	-	-
PS8	16.55	10.3	-	36.5	96.8	511	2.82	-
PS12	4.31	9.3	-	30.4	92.3	390	3.06	-
PS13	11.53	8.3	4.7	32.1	94.8	389	2.58	3.6
CS16	8.34	11.8	1.2	22.1	67.2	363	5.3	3.4
CS17	-	-	10.9	-	-	-	-	2.9
PS20	-	-	3.5	-	-	-	-	4.7
PS23	17.1	8.3	-	-	96	340	-	-
CS24	8.85	9.8	22.9	30	86.5	259	3.27	-
PS26	12	7.1	-	34	93	-	2.24	-
PC27	9.63	8.2	-	34.2	80	227	3	-
CC28	13.3	7.2	-	32.3	81	567	2.79	-
PC29	6.7	7.7	-	34.1	82	380	2.74	-
CC32	52.8	7.2	-	35.7	93	357	2.21	-
CC33	15.1	4.8	-	31.2	74	113	2.08	-
CC35	15.9	5.1	-	30.4	76	427	2.21	-
CC37	18.3	7.6	-	30.8	90	259	2.72	-
CC38	21	7.4	-	27.8	83	224	3.2	-
PC40	9	6.5	-	41.7	87	462	1.72	-
PC41	12.8	7.12	-	34.2	90	211	2.9	-
PC42	19.5	7	-	33.5	79	496	2.62	-
CC43	9.05	7.2	-	34.7	100	379	2.08	-
CC44	15.1	7.3	17.6		70	391	-	-
PC46	7.88	7.7	-	32.8	82	400	2.85	-
PL47	12.2	7	-	29.6	111.6	646	2.42	-
CL50	5.8	5.6	-	26	132	132	2.84	-
CL55	-	6.5	-	-	-	-	-	-

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