SICKLE CELL DISEASE AWARENESS AMONGST COLLEGE STUDENTS

BY

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THESIS

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This descriptive study was designed to investigate if college students attending a midwestern university are aware of the clinical manifestations, treatments, and genetic counseling methods for sickle cell disease. This study was also devised to determine whether or not students, who are more likely to be genetically affected by sickle cell disease, are more or less aware of their sickle cell disease status. Two hundred and fifty-nine (259) University of Illinois Urbana- Champaign (UIUC) students, 18 years and older, enrolled in one of the three following Community Health courses: Community Health Organizations (CHLH 210), Health Care Systems (CHLH 250), and Introduction to Medical Ethics (CHLH 260) were used as study participants. These 259 participants were assessed on their general knowledge of sickle cell disease (SCD). Participants in this study were given a sickle cell disease questionnaire that consisted of 11 questions on sickle cell incidence, prevalence, origin, counseling methods, and knowledge of trait status. Frequency tables, cross-tabulations, and chi-square tests were used to evaluate the variations of existing SCD knowledge among students. Results illustrated that participants did have some general knowledge of sickle cell disease. Study results showed a statistical difference in the response rates for males and females when surveyed on the life expectancy of sickle cell disease (p = .047). Other results showed no statistical differences in response rates between ethnicities group and age.
To my grandfather, who truly believed I could fly.
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CHAPTER 1
INTRODUCTION

1.1 Background

Sickle cell disease (SCD) is an inherited blood disorder caused by abnormal hemoglobin (Creary, Williamson, & Kulkarni, 2007). Sickle cell disease limits the oxygenating role of hemoglobin, resulting in the damaging or the “sickling” of the red blood cells (Barakat, Schwartz, Simon, & Radcliffe, 2008). This disorder affects all parts of the human body and differs widely among individuals (Bloom, 1995). In 1910, Dr. James Herrick, a Chicago physician, was the first American to formally report and identify elongated, sickle-shaped hemoglobin in an anemic Grenadian student’s blood smear. Herrick coined the now familiar term “sickle cell” (Ogamdi, 1994). The sickle-shaped red blood cells described by Herrick caused several complications, including chronic anemia, vaso-occlusive pain episodes, ischemic organ damage, infections, small stature, and delayed puberty (Barakat et al., 2008). For many generations sickle cell disease has been a prevalent disorder in Africa. Reports show that sickle cell disease was a well-known disorder in West Africa and that the West African natives had several local names for this disease before it was discovered in America (Reid & Rodgers, 2007).

Sickle cell disease affects millions of people throughout the world, and it is found to be the most common blood disorder among families whose ancestors came from Sub-Saharan Africa, South America, Cuba, Central America, Saudi Arabia, India, and the Mediterranean regions (Creary et al., 2007). Studies indicate that approximately 1 in 12 African-Americans are heterozygous for the disorder, and approximately 1 in 500
African-American newborns are diagnosed annually with SCD (Boyd, Watkins, Price, Fleming, & DeBaun, 2005). Also, the life expectancy for SCD has doubled since the 1960s. Before that time, few patients lived to reach adulthood (Platt, Brambilla, Rosse, Milner, Castro, Steinberg, & Klug, 1994).

It was not until the 1970s that this blood disorder began to capture public attention in the United States. Prior to that time, many researchers held numerous misconceptions about the nature and course of the disease. Richard Nixon was the first president to make sickle cell disease a matter of national concern by signing the Sickle Cell Anemia Control Act of 1972 (Cerami, 1974). In 1971, President Nixon focused his health message to Congress on sickle cell disease, which at that time was a virtually unknown inherited blood disorder in the African-American community (Reid & Rodgers, 2007). The 1972 act set the foundation for funding toward sickle cell screenings, counseling programs, and the development and distribution of sickle cell anemia educational materials to the general public (Woolley & Gerhard, 1999). With the help of President Nixon, several sickle cell disease research organizations were created, such as the Sickle Cell Disease Association of America (SCDAA), which was established by Charles Whitten in 1972. The SCDAA was designed to improve the quality of life for patients and families with sickle cell disease (Reid & Rodgers, 2007).

After the 1970s, the public’s focus unfortunately shifted once again. The new law, which first established sickle cell education, genetic screenings, and counseling, was stated to be “fraught with controversy” (Treadwell, McClough, & Vichinsky, 2006).
Even the African-American community, which has a higher probability of inheriting SCD, began to regard informed reproductive decision methods, such as screening and counseling, with trepidation and distrust (Treadwell et al., 2006). After President Nixon turned sickle cell disease into a national priority, legislators quickly began to pass laws that mandated premarital and pre-school screenings for sickle cell disease. The U.S Air Force began to deny airmen, who were diagnosed as carrying the sickle cell trait, occupational opportunities if they were applying to be pilots or co-pilots, and insurance companies even increased premiums for individuals with the trait (Reid & Rodgers, 2007). The American people began to view these new legislative policies as genocidal, and these policies were eventually overturned (Reid & Rodgers, 2007).

The lack of national concern for SCD created a barrier in health care. Complications due to the sickling of the red blood cells therefore continue to be a significant issue to patients and physicians in today’s medical world. Physicians remain puzzled by the biological and clinical intricacies of SCD, and SCD researchers are trying to find a cure to reverse the “sickling effect” in the human body.

1.2 Purpose

Sickle cell disease continues to be a global health problem that presents major challenges to our health care systems. The reviewed SCD literature expresses a dire need for more public education and awareness on SCD in the United States. In comparison with other chronic diseases and blood disorders, sickle cell disease remains one of the least understood and puzzling medical conditions by health care workers and the general
public, as well as the least funded blood disorder (Clarke & Clare, 1981). Misleading descriptions of sickle cell disease, as a race-related disease during the past few decades, have significantly contributed to the rise in the public’s misunderstanding of sickle cell disease (Clarke & Clare, 1981).

In addition, existing research on SCD focuses on the awareness of the disease among college students attending Historically Black Colleges and Universities (HBCUs) where the majority of participants are of African-American descent. Therefore, a paucity of information exists regarding the awareness among college students attending midwestern universities. This study therefore attempts to determine if college students attending a midwestern university are aware of the clinical manifestations, treatments, and genetic counseling methods for sickle cell disease. This study hopes to determine whether or not students, who are more likely to be genetically affected by this disease, are more or less aware of their SCD status.

1.3 Research Questions

This research study attempts to answer the following three questions by using a sickle cell disease questionnaire to survey college students on their existing knowledge of sickle cell disease.

Research Question 1

How knowledgeable are midwestern college students on background information regarding sickle cell disease?
Research Question 2

Do any significant differences exist in awareness between ethnic groups or groups of students who have a higher probability of inheriting sickle cell disease traits?

Research Question 3

Do any significant differences exist in awareness between gender groups?

1.4 Definitions

For the purpose of this literature review, these phrases, frequently referenced throughout the text, are defined as follows:

- *Genetic counseling*: Communication process between health care provider and client that emphasizes and provides accurate and up-to-date information about a genetic disorder in a sensitive and supportive, non-directive manner (SCDA, 2005).

- *Hemoglobin*: Chemical substance (an iron containing protein) of the red blood cell, which carries oxygen to the tissues, and gives the cell its red color (SCDA, 2005).


- *Hemoglobin C trait* (AC): Inheritance of one gene for the usual hemoglobin (A) and one gene for hemoglobin (C). A person who has the hemoglobin C Trait (AC)
is a carrier of the hemoglobin C gene, and is not affected by the gene (SCDAA, 2005).

- **Hemoglobin C disease**: A person has both HbS and HbC and is often referred to as “HbSC.” Hemoglobin C causes red blood cells to develop. Having just some hemoglobin C and normal hemoglobin, a person will not have any symptoms of anemia. However, if the sickle hemoglobin S is combined with the target cell, some mild to moderate anemia may occur (UMMC, 2010).

- **Hemoglobin E disease**: Similar to sickle cell-C disease except that an element has been replaced in the hemoglobin molecule under certain conditions, such as exhaustion, hypoxia, severe infection, and/or iron deficiency (UMMC, 2010).

- **Hemoglobin S-beta-thalassemia**: An inheritance of both the thalassemia and sickle cell genes. The disorder produces symptoms of moderate anemia and many of the same conditions associated with sickle cell disease, but to a milder degree (UMMC, 2010).

- **Sickle cell anemia (SS)**: An inherited disorder of the red blood cells in which the hemoglobin is different from the normal hemoglobin. This unusual hemoglobin results in the production of unusually shaped cells and is referred to as “HbSS.” It is the most common and severe form of the sickle cell variations (SCDAA, 2005).

- **Sickle cell disease (SCD)**: An inherited disorder of the red blood cells in which one gene is for sickle hemoglobin (S) and the other gene is for unusual hemoglobin such as S, C, Thal (SCDAA, 2005; UMMC, 2010).
• **Sickle cell trait**: A person carrying the defective gene, HbS, but also has some normal hemoglobin HbA. Persons with the sickle cell trait are usually without symptoms of the disease, but mild anemia may occur under intense, stressful conditions, exhaustion, hypoxia (low oxygen), and/or severe infection. The sickling of the defective hemoglobin may occur and result in some complications associated with sickle cell disease (UMMC, 2010).

**1.5 Assumptions**

The following two assumptions have been made: 1) The data are reliable and accurate measurements of a student’s awareness, and 2) each participant answered the questionnaire without coercion.
CHAPTER 2

LITERATURE REVIEW

Introduction

The review of literature is a comprehensive overview of sickle cell disease. The following review includes background information on sickle cell disease, as well as information on the following topics: complications, treatments, federal government initiatives to support and improve sickle cell disease awareness in the United States, newborn screening, genetic testing, the patient-physician medical relationship, and the perception of sickle cell disease among health care providers. The primary goal of this review is to highlight the lack of extensive knowledge and awareness of sickle cell disease among sickle cell disease patients, providers, and the general public.

2.1 Natural History/Natural Selection

Historians believe that DNA mutations, which were responsible for the first versions of the sickle cell gene, originally arose in various African regions (see Figure 2-1), including Cameroon, Central Africa Republic, Benin, and Senegal (Jones, 2008). Studies show that the Trans-Atlantic slave trade introduced the sickle cell gene into the Americas and the Caribbean islands (see Figure 2-1). African slaves, who carried the sickle cell trait, had the specific β-globin gene. Popmpa (1996) showed that carriers, who had the sickle cell trait, had a heterozygote advantage of being resistant to malarial
infection. The Indo-European sickle mutation also originated in the Indus Valley Harappa. This particular mutation is found in Saudi Arabia, Oman, Bahrain, and Kuwait.

Figure 2-1: Geographical distribution of the sickle gene (Stuart & Nagel, 2004).

A British colonial medical officer, E. A. Beet, stationed in Zimbabwe in the 1940s, first observed that blood from malaria patients, who carried the sickle cell trait, had fewer malarial parasites compared to blood from patients without the trait (Bloom, 1995). However, in the 1950s, Anthony C. Allison developed his own hypothesis on the association of malaria and the sickle cell trait. By following up on Beet and a physician in Zaire during that time, Allison was able to hypothesize that the sickle cell trait offered protection against malaria. He believed that people with the sickle cell trait did not easily succumb to malaria as often as people without the trait (Bloom, 1995). Some evidence against Allison’s hypothesis shows no difference in the concentration of blood-borne malarial parasites that exist in people with the sickle cell trait compared those without it.
(Bloom, 1995). However, research shows that the sickle cell traits offer more protection against malaria to children than to adults, and adults are able to develop antibodies that can attack parasites in the immune system and increase their survival rate in malarial climates. Compared with adults, young children are not able to produce antibodies to the malaria disease until their immune system is more mature (Bloom, 1995).

2.2 Genetics

Sickle cell disease is one of the many hemoglobin variants that cause mutations to alter amino acids in the hemoglobin molecule (Popma, 1996). Normal adult hemoglobin HbA is a heterogeneous mixture of approximately 90% Hb A, 2.5% Hb A2, 3.5% Hb Alc, and small quantities of fetal hemoglobin (Popma, 1996). All normal hemoglobin molecules consist of four polypeptide chains in which two are globin \( \beta \) chains and \( \alpha \) globin chains (Popma, 1996). Sickle cell disease occurs when the hemoglobin produced is HbS instead of HbA. Genetically, sickle cell disease occurs when valine replaces glutamic acid at position 6 of the \( \beta \) globin chain (Popma, 1996).

In the United States, three prevalent genotypes of sickle cell disease occur (see Table 1). The genotypes are abnormal hemoglobins designated by their mutations within the globin chain. Homozygous SCD, HbSS, or sickle cell anemia, is known as one of the more common types of sickle cell beta genes inherited by both parents (SCDAA, 2005). Sickle cell hemoglobin SC is known as one of the milder traits formed by one S-globin chain and one \( \beta \)-globin chain. This gene is found among West Africans (Bloom, 1995). Also, the S\( \beta^0 \)-thalassemia and HbSS are classified as being more severe compared with
other genotypes, and they are found among populations from the Mediterranean region (Bloom, 1995).

2.3 Inheritance Patterns

Sickle cell disease is a recessive gene if two parents have two copies of the Hb S gene. Children born to parents with these genes (see Figure 2-2) will have sickle cell disease.

Figure 2-2: Two parents with sickle cell disease (Bloom, 1995).

2.3.1 One Parent has Sickle Cell Disease and Sickle Cell Trait

Figure 2-3 shows that one parent has the sickle cell disease Hb S genes; therefore, all of that parent’s gametes will carry the Hb S gene. The other parent has one Hb S gene and one Hb A gene. The chances for that parent with both the Hb S gene and the Hb A gene transmitting either gene are equal, or 50/50 (Bloom, 1995). All children born to
these parents will have either sickle cell disease or sickle cell trait children with the chances being 50/50 for each (see Figure 2-3).

Figure 2-3: One parent has sickle cell disease and sickle cell trait (Bloom, 1995).

2.3.2 One parent has sickle cell disease and the other parent carries normal genes

Figure 2-4 illustrates that the Hb A gene is carried by the normal parent and the gametes from this parent will carry this particular gene, as well as the sickle cell disease parent carrying the Hb S gene (Bloom, 1995). All children conceived from these parents will inherit one normal and one sickle cell gene. All children will have the sickle cell trait.

Figure 2-4: One parent has sickle cell disease and the other parent carries normal genes (Bloom, 1995).
2.3.3 Two parents has sickle cell trait

This case (see Figure 2-5) illustrates that both parents have an equal chance of transmitting the two genes. If both parents have the sickle cell trait, they will have a 25% chance of having children with sickle cell disease, as well as a 50% chance of carrying the sickle cell trait (Bloom, 1995). Also, a slight 25% chance occurs of children with normal genes. In this case, 50% of the time one parent will transmit an Hb S gene, and the other half of the time the other parent will transmit an HbA gene (Bloom, 1995).

![Figure 2-5: Two Parents has Sickle Cell Trait (Bloom, 1995).](image)

2.3.4 One parent has sickle cell trait and the other is normal

All children from this couple (see Figure 2-6) will display the sickle cell trait or have normal genes because half of the gametes of the parent who carries the sickle cell trait will carry the Hb S gene, and the other half will carry the Hb A (Bloom, 1995). This combination will result in a 50/50 chance of producing a child with normal genes or inheriting sickle cell traits.
Figure 2-6: One parent has the sickle cell trait and the other is normal (Bloom, 1995).

2.4 Complications

Vaso-occlusive crises, or pain episodes, are severe and complicated forms of sickle cell disease that occur in all parts of the body. According to Marlowe and Chicella (2002), vaso-oclusive episodes have been repeatedly reported as the most common cause for hospitalization of SCD patients, representing $475 million in annual health care expenditures and $75,000 in hospitalization costs. Adolescents and adults who suffer from vaso-occlusive crises experience severe and recurrent pain episodes throughout their lifetimes. The two types of vaso-occlusive pain are acute and chronic episodes. Acute vaso-occlusive episodes involve pain for periods of less than three to six months (Payne, 1989).

Marlowe and Chicella (2002) noted that acute sickle cell pain has been described by patients as being more severe than post-operative pain and as intense as cancer pain. Pain episodes can begin as early as 6 to 9 months of age and continue throughout adulthood, with episodes lasting for days or even weeks (Steinberg, 1999). Recent studies suggest that children born with the SS genotype have a significantly median age
of 1.9 years to first dactylitis episodes, compared to 3.9 years with those who are born with the SC genotype (Dampier, 2008). Also in terms of episodes, 40% to 50% of those children experience one pain episode per month, while 10% of other school-age children experience pain episodes more than twice a month (Dampier, 2008).

Unfortunately, the underlying mechanism responsible for the onset of pain episodes remains ambiguous to researchers and experts who study SCD. Researchers believe that the onset of pain episodes can be provoked by changes in altitude, temperature, physical and emotional stress, menstruation, fatigue, and dehydration (Marlowe & Chicella, 2002). Steinberg (1999) indicated that, in a given year, up to 60% of patients with SCD experience severe pain episodes, with a small majority of patients suffering from severe pain almost constantly.

Some patients with SCD require frequent hospitalization, while others seldom experience episodic pain (Gil, Abrams, Phillips & Keefe, 1989). In addition, people with a range of mild to severe symptoms of SCD often complain of pain in four major parts of the body: the abdomen, low back, joints, and chest (Gil et al., 1989). Dampier’s (2008) research illustrated that the impact of pain episodes goes beyond the substantial use of health care resources. The impact of pain episodes has shown to have adverse effects on children’s physical functioning, school attendance, academic performances, and social roles.

The management and prevention of pain episodes continue to be a major challenge for health care professionals and researchers. Although no known cure exists
for sickle cell disease, we can have room for improvement in the care process for sickle cell disease patients in the United States. Prabhakar (2009) noted a lack of thorough understanding of the pathophysiology of sickle cell disease, as well as observing that the primary focus of SCD clinical care revolves around treating the complications of SCD rather than focusing on the prevention of complications. Pain from vaso-occlusive crises is the most common clinical problem experienced by patients with SCD, and often necessitates emergency visits to hospitals.

Much of the literature has confirmed that the most effective method of managing pain is to treat the underlying medical condition, which includes minimizing major factors associated with acute intravascular sickling (Payne, 1989). Drug therapy continues to be the primary form of treatment for acute and chronic SCD pain. Major medical controversies regarding drug treatment include the use of narcotic analgesics for the management of SCD pain and the incidence of substance abuse among SCD patients treated with narcotic analgesics for pain (Payne, 1989).

2.4.1 Stroke

A childhood stroke is one of the many devastating complications caused by sickle cell disease that can result in permanent brain damage or death. A stroke is triggered by a decrease in the flow of oxygen due to a major disruption of the amount of oxygen to the brain. This complication can occur without warning. Studies have found that the median age for a stroke in SCD is 6 years of age. Studies have also revealed that by the age of 14, a stroke has affected at least 8% of the SCD patient population, and by the age of 20, a
stroke has affected 11% of the patient population (Njamnshi, Mbong, Wonkam, Ongolo-Zogo, Djientcheu, Sunjohy, 2006; Serjeant, 1997). The reoccurrence rate for strokes for children within three years of SCD diagnosis is 50% to 70% (Serjeant, 1997).

Transcranial Doppler Ultrasonography (TCD) and Magnetic Resonance Imaging (MRI) can be used to diagnose strokes. The TCD predicts the levels of risk for strokes by measuring mean blood velocity (Mazumdar, Heeney, Sox, & Lieu, 2007). The existing literature indicates a need for a more accurate diagnostic method for strokes other than the use of TCD. In addition, several research studies have shown a lack of educational materials about the effects of strokes, and these studies indicate a need for more stroke screening tests. The extent of this problem was revealed in a cross-sectional study conducted by The Children’s Hospital of Philadelphia (CHOP) using a population of 44 children with sickle cell disease and 50 caregivers (Katz, Whitley-Smith, & Frempong-Ohene, 2002). The research showed that 46% of the caregivers surveyed were unable to identify the warning signs of a stroke. In this study, only 34% of the caregivers could identify a stroke as being a complication associated with SCD (Katz et al., 2002).

### 2.4.2 Acute Chest Syndrome

Acute Chest Syndrome (ACS) is a serious complication of sickle cell disease that contributes to the high rates of morbidity and mortality in SCD. The ACS results from infections in the lung cavity caused by bacteria organisms. The ACS is a serious complication similar to pneumonia, and has been reported to occur in about 50% of sickle
cell disease patients; ACS has reoccurred in approximately 80% of SCD individuals (Fawibe, 2008).

The Acute Chest Syndrome ACS accounts for approximately 25% of premature deaths among SCD patients. No available laboratory, radiographic, or clinical methods accurately establish the etiology of ACS. Researchers argue that infection and fat emboli are the most readily identifiable causes of ACS (Bernard, Yasin, & Venkat, 2007; Fawibe 2008). The most common diagnostic symptoms in all age groups are fever, coughs, abnormal chest x-rays, and chest pains. Less common symptoms include shortness of breath, productive cough, and wheezing (Bernard et al., 2007).

2.5 Treatments

Hydroxyurea is a popular cytostatic myelosuppressive, chemotherapeutic agent drug approved by the FDA in 1998 for adult sickle cell disease patients (Weiner & Brugnara, 2003). For many decades, hydroxyurea was used as a chemotherapeutic drug to treat cancer. Hydroxyurea is currently used to decrease sickle cell hemoglobin polymerization and erythrocyte sickling by increasing fetal hemoglobin (HbF) production, a hemoglobin found in newborn babies (Weiner & Brugnara, 2003). This drug has successfully reduced the number of painful periods and the recurrence of prevalent SCD complications. Physicians recommend that individuals who experience frequent painful episodes or who have a history of acute chest syndrome, symptomatic anemia, and other vaso-occlusive episodes are candidates for hydroxyurea therapy (Mehta, Annan-Afenyi, Byrns, & Lottenberg, 2006).
Adults in a random placebo-controlled trial study with hemoglobin SS, who were receiving hydroxyurea, exhibited a significant reduction in vaso-occlusive crises and the incidence of acute chest syndrome as a result of hydroxyurea therapy (Mehta et al., 2006). In this study, painful episodes decreased from 4.5 to 2.5 painful crises per year (Mehta et al., 2006). A similar double-blind study, which involved placebo-controlled trials of hydroxyurea with 299 adult SCD participants, terminated after 28 months of treatment because hydroxyurea reduced the frequency of pain episodes in patients along with the frequency of episodes of acute chest syndrome (Steinberg, 1999).

The U.S. Food and Drug Administration (FDA) has not approved hydroxyurea treatment for use by infants and children. Studies of hydroxyurea in young children are in the early stages, and long-term follow-ups need to ascertain the risk of continuing adverse side effects on children (Mehta et al., 2006). A pilot study of hydroxyurea by the National Heart, Lung, and Blood Institute (NHLBI) evaluated whether or not hydroxyurea can benefit young patients. Participants who ranged from 6 to 24 months of age demonstrated that the drug is well tolerated in younger children. Scientists predict that this drug will be an important factor in emergent countries. Many developing countries do not have the facilities necessary to safely perform bone marrow transplants and transfusions.

Hydroxyurea has been described as a cost-effective treatment. Moore, Charache, Terrin, Barton, & Ballas (2000) examined the cost-effectiveness of hydroxyurea therapy and found a total cost savings of almost $26 million a year if every eligible patient in the
United States were taking hydroxyurea (Lanzkron, Haywood, Hassell, & Rand, 2008; Moore et al., 2000). Moore et al. (2000) study results showed (see Table 2) that hydroxyurea averaged $16,810 for the annual medical care cost for patients while those cost of control patients averaged $22,020 (Nietert, Silverstein, & Abboud, 2002).

2.5.1 Over-the-counter Drugs

Over-the-counter medicines, such as nonsteroidal anti-inflammatory drugs NSAIDs, acetaminophen (Tylenol), ibuprofen (Advil or Motrin), as well as opioids, such as codeine, morphine, and oxycodone, are used to control sickle cell pain. Non-opioids are prostaglandin-synthesis inhibitors, prescribed for the management of mild pain and used in conjunction with opioids (Stinson & Naser, 2003). Non-opioids prevent the conversion of arachidonic acid to prostaglandins by interfering with the function of the enzyme cyclo-oxygenase (COX) (Stinson & Naser, 2003). Non-opioids can provide additive analgesia when combined with opioids. Acetaminophen is another popular drug use for the treatment of sickle cell disease. Acetaminophen is described as one of the safest analgesics, and it inhibits the synthesis of prostaglandins. On the other hand, NSAIDs are prescribed for patients with gastritis, peptic ulcer, and renal failures. Opioids are a popular analgesic used for the treatment of moderate and severe acute pain due to pain episodes (Stinson & Naser, 2003).

2.5.2 Transfusion Therapy

Transfusions are another method of trying to deal with SCD complications and assisting with the prevention of strokes and acute chest syndrome. Transfusions are
recommended for use of SCD patients with acute chest syndrome, heart failure, and severe anemia (in children) who have enlarged spleens, multi-organ failure syndrome, splenic sequestration, aplastic crisis, and who are susceptible to strokes (Claster & Vichinsky, 2003). Transfusion therapy seeks to increase the concentration of hemoglobin A and reduce the percentage of sickle hemoglobin to increase the oxygen carrying capacity of blood (Claster & Vichinsky, 2003).

A clinical trial study, which evaluated the efficacy of transfusions among patients with some SCD complications, showed that repeated transfusions reduce the risk of recurrent strokes in children with SCD. This study also showed that researchers were able to predict that 50% of children with SCD, who had suffered strokes and had not received transfusions, would suffer strokes within three years compared to 10% of SCD individuals who had received transfusions (Steinberg, 1999).

2.5.3 Bone Marrow Transplantation

Bone marrow transplantation is an experimental therapy limited to candidates below 16 years of age and who exhibit SCD complications, such as a stroke, acute chest syndrome, and refractory pain (Steinberg, 1999). Statistics show that only 1% of patients with sickle cell anemia actually meet the set requirements for bone marrow transplantation (Steinberg, 1999). In 1984, a bone marrow transplant was used to treat a pediatric patient with leukemia. The transplant also cured the patient’s sickle cell disease (NIH, 2002).

Researchers have found that the mortality rate from this procedure is 10% and that it requires SCD patients to undergo a chemotherapeutic regimen prior to transplant.
Consequently, some SCD patients do not survive the chemotherapy process whereas others may suffer from life-threatening infections before their bone marrow and immune systems are sufficiently regenerated (NIH, 2002). Potential complications include graft rejection and graft versus host disease (GVHD). Currently, transplants are still considered only for sickle cell disease patients whose disease is severe enough to justify the risk. The National Heart, Lung, and Blood Institute (2002) is looking for various methods to reduce the risk of bone marrow transplantation. The blood from a baby’s umbilical cord is believed to reduce the risks of rejection and graft-versus-host disease. Also, this blood does not need to match the recipient’s blood as closely as bone marrow donors (NHLBI, 2002).

2.6 Experimental Treatment

Gene therapy is a new treatment for patients with sickle cell anemia. Researchers believe that gene therapy is an improvement over blood transfusion because it offers a more permanent solution. Gene treatment requires a delivery system that can carry a normal gene to cells that contain defective genes (Jones, 2008). Scientists have chosen modified viruses as an efficient delivery system. Normal genes are delivered to the target cells to ensure that the normal genes function as they should (Jones, 2008). Gene therapy research is still in an early stage, and many scientists believe that this method could be the clinical cure for sickle cell anemia.
2.7 Government Initiatives

The 1972 Sickle Cell Disease Control Act was the first law to mandate funding for scientific research programs on sickle cell disease. Such programs were designed to improve the quality of life and care for patients with sickle cell disease. These programs aimed at reducing the percentage of deaths from sickle cell anemia by promoting research on treatment and prevention (Bonds, 2005; Jones, 2008).

2.7.1 Sickle Cell Treatment Act

President George W. Bush signed the bipartisan Sickle Cell Treatment Act into law in 2003. This law granted states to get federal funding for patient counseling, educational initiatives, and community outreach programs (Haywood, Hassell, & Rand, 2008; SCDAA, 2005). According to the Sickle Cell Disease Association of America, patients can receive federal matching funds for sickle cell disease-related services under Medicaid.

2.7.2 United States Postal Service

In 2004, the U.S. Postal Service issued a stamp to promote sickle cell disease awareness. The image, created by artist James Gurney, depicts a mother holding her baby. The stamps have an inscription: “Test early for sickle cell” (Jones, 2008). The stamp was created to emphasize the importance of early SCD testing, and it provided the U.S Postal Service with a means of raising public awareness on sickle cell disease (Jones, 2008).
2.8 Newborn Screenings

Newborn screenings are believed to be the most effective and efficient screening programs for the detection and diagnosis of inherited diseases (Bioethics, 2001). Newborn screening programs were designed to help physicians identify newborns with SCD and to allocate penicillin prophylaxis to help reduce the incidence of pneumococcal sepsis in infants. Before government laws mandated the screening of newborns for SCD, the rate of mortality approached 30% by age 3, and most deaths were due to sepsis and acute anemic events (Reed & Vichinsky, 1998).

Early screening for the sickle cell trait and treatment for sickle cell disease can substantially reduce the risk of serious infection during the early years of life (U.S. Preventive Services Task Force, 2007). Follow-up sickle cell screening and counseling have shown to be the most effective methods of helping families with infants who have been identified as having the sickle cell trait. Although all states now screen for sickle cell disease, this disease remains less familiar to the public compared to other blood disorders found by early screenings, such as phenylketonuria (PKU) and hypothyroidism (Yang, Andrews, Peterson, Arvind, & Cepeda, 2000).

2.9 Genetic Counseling

Parents who receive genetic counseling and support learn how to assess respiratory distress, fever, jaundice, and splenomegaly, which are early major complications of SCD. (Claster & Vichinsky, 2003). Research has confirmed that the follow-up genetic counseling rates for infants with sickle cell disease and other
hemoglobinopathies ranges from 35% to 60% nationwide. These low rates indicate that many families are not complying with follow-up procedures for infants with sickle cell disease.

Cultural views, social and economic barriers, guilt, and health beliefs may contribute to the lack of interest and support for sickle cell testing and genetic counseling (Gustafson, Gettig, Morse, Krishnamurti, & Lakshmanan 2007; Yang, Andrews, Peterson, Arvind, & Cepeda, 2000). Articles on prenatal sickle cell screening education suggest that many factors contribute to the lack of follow-up counseling and screening. These factors include anticipatory anxiety on the parents’ part about learning that their infant has a sickle cell hemoglobinopathy, denying and avoiding the possibility that their child might be born with sickle cell disease, and fear of being responsible for causing the disease (Yang et al., 2000). In addition, Hill (2001), the author of Care Giving in African-American Families: Caring for Children with Sickle Cell Diseases, discussed that fathers are usually less likely to know they are carriers of the sickle cell trait and are more than likely to deny their contribution to passing on the disease.

The lack of awareness of newborn screening programs is another key factor that contributes to low follow-up rates. The follow-up rate for newborn screenings may increase significantly only if the parents of infants with sickle cell hemoglobinopathies become informed and aware of the significance of the screening program during prenatal education sessions (Yang, Andrews, Peterson, Arvind, & Cepeda, 2000). Hill (2001) stated that many mothers experience feelings of helplessness and fear because of the SCD
diagnosis. Hill (2001) concluded that mothers receive little information from physicians regarding how to care for their SCD children. This information further demonstrates a lack in awareness of SCD as a result of poor patient-physician relationships.

### 2.10 Patient and Physician Relationship

It is particularly important for sickle cell disease patients to develop trust in their medical professionals, especially those who underutilize recommended medical therapies (Haywood, Lanzkron, Ratanawongsa, Bediako, Lattimer, & Powe, 2010). Sickle cell disease patients develop a sense of mistrust for clinicians for various reasons: not being considered as partners in the medical relationship, negligence while being hospitalized, and doubting the legitimacy of SCD pain (Haywood et al., 2010). Haywood et al. (2010) noted that adults with sickle cell disease are likely to be more aware of their illness than some health care workers, yet many patients have reported in qualitative studies that their acute health care providers do not have their best interest at heart and fail to prescribe treatment applicable to their medical needs.

In addition, studies that have observed the physician communication behaviors between African-Americans conclude that physicians are shown to exhibit less empathy, courtesy, and attention to African-American patients (Haywood et al., 2010). In one study, Haywood et al. (2010) also noted that physicians rated African-American patients more negatively when scoring on intelligence, education level, and the likelihood of not to complying with medical advice, as well as abusing medication. Haywood et al.’s (2010) study on the results of the association of medical provider communication and
trust among sickle cell disease patients’ showed an association between the health care provider relationship and sickle cell disease patient’s trust. Ninety-five adults with sickle cell disease were tested in this study. This study illustrated a 3.76% increase in trust scores from sickle cell disease patients when there was a 10% increase in provider communication ratings (Haywood et al., 2010). Researchers therefore concluded that 1) a poorer patient rating of provider communication will result in a decrease in trust among patients, and 2) communication has a direct effect on a patient’s receptiveness to his/her physician (Haywood et al., 2010).

2.11 Health Care Professionals’ Perceptions and Knowledge of SCD

For patients to receive quality and efficient care, it is imperative that health care professionals and caregivers are aware of how to treat SCD. A study at Southern University Teaching Hospital in Baton Rouge, Louisiana, showed that health care workers, especially nurses, have major difficulty treating patients with chronic illnesses, such as sickle cell disease, because SCD patients do not fit into the preconceived “sick role” found among hospital patients with acute illnesses (Mabien, Labbe, Herbert, & Haynes, 2001). Nurses have been reported to assess SCD patients’ sickle cell pain less seriously than for patients who do not have SCD (Mabien et al., 2001).

The preconceptions of health care professionals regarding SCD results from a lack of understanding of how to treat patients coupled with the fear of creating pain killer addicts when distributing medicine to SCD patients (Mabien et al., 2001). Health care professionals continue to mistreat SCD patients, particularly minorities, by withholding
the requisite analgesics. Chestnut (1994) challenged the issue of race and gender in his Service Perception Test (SPT), a pilot study that examined how ethno-cultural factors of age, race, and gender were perceived as influencing the quality of health care received by patients.

Chestnut’s (1994) study results showed, without fail, that 1) white patients were perceived to get better sickle cell care than blacks, 2) young children were provided with better care than the elderly, and 3) the elderly were provided with better care than middle-aged adults (Prabhakar, 2009). Moreover, when SCD patients remain untreated, studies show that they are predisposed to developing pseudo-addictive behavior (Marlow & Chicella, 2002), which causes patients to seek and hoard drugs due to the fear of future pain. Pseudo-addictive behavior ends when patient pain is properly managed and patients receive the proper dosage of medication. Sickle cell disease and pain management literature dispelled several myths about SCD. Studies have shown that SCD patients are no more susceptible to developing an addiction to pain medication than any other group of patients (Jacob, 2006). A patient care study conducted at Southern University Teaching Hospital, using a sample composed of licensed practical nurses, pediatric nurses, and registered nurses, showed a lack of empathy for SCD patients on their part. This study revealed a lack of understanding of SCD and pain for SCD patients (Mabien. Labbe, Herbert, & Haynes, 2001). Health care providers managed to show some level of sympathy for patients with cancer pain, post-operative pain, and pain due to trauma. On the other hand, providers still found it challenging to sympathize with people who experienced SCD pain (Mabien et al., 2001).
2.12 Sickle Cell Knowledge

During the past four decades, the federal government has made significant strides in providing funding and research on SCD. However, much of the reviewed literature shows a lack of knowledge on SCD among at-risk populations. In a study by Boyd et al. (2005), 264 African-American women between the ages of 18 and 30 from St. Louis, Missouri, participated in a cross-sectional telephone survey on SCD. These authors found that 30% of the African-Americans contacted had no prior knowledge of SCD and were released from their study. Of the 162 women who met the eligibility criteria, only 9.3% understood the inheritance pattern of SCD, and 11% were unaware of their carrier status (Boyd et al., 2005).

In addition, participants did not understand treatment strategies for SCD, yet most participants were well aware that SCD is a defective blood disorder and that pain episodes are a major complication (Boyd et al., 2005). This study provided strong evidence that African-American women in their prime reproductive age are still not equipped with adequate information on SCD, SCD incidence, and inheritance patterns (Boyd et al., 2005). Such general information is essential when making informed decisions on having a child (Boyd et al., 2005).

Treadwell et al. (2006) surveyed 282 people from northern California about their exposure to and knowledge of sickle cell disease and sickle cell traits. Interestingly, 68% of those interviewed in this study responded correctly to knowledge questions about SCD. Only 15% of the respondents were aware of their own trait status. A majority of
respondents reported that they received SCD testing at their local community, hospital, and clinic (Treadwell et al., 2006).

Ogamdi (1994) evaluated the general knowledge of SCD among 334 students from a university in southeastern Texas. Study results illustrated that approximately 81% of students were not unaware of the genotype describing SCD, and more than 60% of students were unaware that SCD is a preventable disease if individuals made “responsible” reproductive decisions (Ogamdi, 1994). On the other hand, 63% of students were able to correctly answer knowledge questions regarding symptoms of SCD. A significant limitation of this study was that not all questionnaire responses were reported in frequencies. However, this researcher has concluded that individuals between the ages of 19 and 30 lack the understanding of basic facts concerning SCD. A need exists for more sickle cell screenings, education, and counseling among university students.

In a similar study, Stewart and colleagues conducted a mixed-method study using a sample of 191 African-American college students from the Southeast who ranged from 19 to 30 years of age (Prabhakar, 2009). Their study focused on examining the knowledge and belief systems surrounding SCD, the SCD trait, and genetic testing among young African-American adults (Prabhakar, 2009). The investigators found a lack of knowledge regarding carrier status, family history, and genetic testing among the target group (Prabhakar, 2009).

Finally, in a research study designed by Dyson (1997), 104 carriers and non-carriers of SCD were evaluated on their knowledge and awareness of SCD. Results
showed that participants poorly understood questions related to patterns of inheritance. Participants answered only 25% and 29% of the inheritance questions correctly. Interestingly, no significant difference occurred in knowledge of SCD among carriers and non-carriers. Both scores were believed to be “strikingly” similar by the researcher (Dyson, 1997). Three carrier participants claimed to receive counseling, but no significant differences occurred between those counseled and those not counseled. This study had several limitations. With 104 participants, only 55 completed the survey. Researchers believed this discrepancy to be a result of incomplete or inaccurate addresses, as well as several methodological oversights.
2.13 Conclusion

Sickle cell disease is a disease that affects more than 100,000 African-Americans in America. After reviewing much of the literature on this subject, this researcher determined that a great need exists for more research and education on this disease, especially for more research studies that examine SCD awareness among various ethnic populations in America. The scarcity of SCD research illustrates how our society fails to view sickle cell disease as a serious illness. Currently, our society’s attention is on non-inherited blood disorders, for example, HIV/AIDS, hypertension, and cancer. Without awareness and a public outcry for a cure and more funding, sickle cell disease will continue to be a silent killer to young men and women around the world.
CHAPTER 3
RESEARCH METHODS

3.1 Participants

Participants in this study were undergraduate students from the University of Illinois, at Urbana-Champaign who were enrolled in one of the three following Community Health courses: Community Health Organizations (CHLH 210), Health Care Systems (CHLH 250), and Introduction to Medical Ethics (CHLH 260). The exclusion criteria for this study included any person not enrolled in the Community Health courses 210, 250, and 260, unwillingness to participate, and students under the age of 18. Two hundred fifty-nine (259) questionnaires were distributed among the three classes. The use of undergraduate participants as the study sample followed the procedures and guidelines set forth by the University of Illinois at Urbana-Champaign and the Institutional Review Board (IRB) (see Appendix C). Participants were not placed in physical, emotional, or academic harm at any time during the course of the study. The study design was reviewed and approved by the IRB.

3.2 Procedures/Materials

The Sickle Cell Disease (SCD) questionnaire was administered to all eligible participants between November and December 2008. Before the questionnaires were distributed, a consent form was read and distributed to all students for their review (see Appendix D). The consent form informed participants that the research study was voluntary and that they would not be placed at any risk following the completion of the
study. After the consent form was read and distributed, students had approximately 10 to 15 minutes to complete their questionnaire in class under the supervision of their professor. To maintain the confidentiality of all participants, names and signatures were not retrieved on the questionnaires. Members of the research team collected the questionnaires to protect the anonymity of participants. All consent materials and questionnaires were stored in a secure office.

3.3 Survey Instrumentation

The instrument used for this study was a structured questionnaire containing multiple choice questions and fill-in-the-blank demographic questions. Section 2 contained 11 multiple-choice questions that tested students’ knowledge of clinical characteristics of sickle cell disease, populations at risk for developing SCD, screening and counseling methods, incidence, and prevalence rates. Two faculty members of the Community Health Department reviewed the questionnaire. Descriptive questionnaire data were processed and analyzed in the Statistical Service Database for Windows (SPSS).

3.4 Study Measure

All questionnaire data were entered into the SPSS database and coded by using a numerical system. The questionnaire contained four categories of race identification for participants to identify: 1) Caucasian, 2) African-American, 3) Hispanic, and 4) Other. Gender was coded as 1) male and 2) female. Age was determined by students filling in the blank. Ten sickle cell disease questions were listed on the questionnaires. Frequency
tables, cross-tabulations, and chi-square tests were used to evaluate the variations of existing SCD knowledge among students and the statistically significant alpha p value ($p = 0.05$) was used to determine variations in response rates. Frequency tables were divided by ethnicity and response results, whereas cross-tabulation tables grouped all response variables together by ethnicity per result question. The chi-square test was used to group students’ response rates by gender and ethnicity.
CHAPTER 4
RESULTS

4.1 Response Rates

Two-hundred fifty-nine questionnaires were distributed to students at the University of Illinois at Urbana-Champaign in three community health courses. Although 259 participants were included in the analysis, not all participants responded to each question. Percentages reported in this study are analyzed results from the total number of participants who responded to each question. Those questions left unanswered were coded as (0) and did not conflict with the overall analysis and study.

4.2 Sample Description

Each participant’s demographic information is shown in Table 5. Of the 259 participants who responded to the questionnaire, approximately 54% of the respondents identified themselves as Caucasian, 22% identified themselves as being African-American, 0.6% identified themselves as Hispanic, and 17% identified themselves as “Other.” The majority of students who identified themselves as “Other” were of Asian and Pacific-Islander descent.

4.2.1 Gender

The average age for participants was 20 years old (see Table 3), and the age range varied between 18 and 29 years of age. Approximately 69.1% of the participants identified themselves as female, 30.1% of the participants identified themselves as male, and 1% of the participants remained unresponsive.
4.3 Knowledge of Sickle Cell Disease

The knowledge questions that focused on sickle cell disease were evaluated by calculating the number of correct and incorrect responses for each participant in SPSS for Windows. Approximately 92.7% of the participants were able to identify sickle cell disease as being an inherited blood disorder. Eighty-one percent of the participants knew that sickle cell disease evolved from Africa. On the other hand, only 17% of the participants were able to identify the number of new births born with sickle cell disease. Interestingly, 84.9% of the participants were able to identify the clinical manifestation of sickle cell disease.

Out of the total number of participants, 94.2% correctly identified that no cure for sickle cell disease exists along with 91.1% of the students correctly identifying populations most at risk of developing sickle cell disease. Only 78.8% of the students could identify that sickle cell anemia is a medical condition in which the red blood cells develop an unusual shape, and 89.2% identified the appropriate time to test for sickle cell disease. Table 8 shows that only 43.6% of the participants were able to identify the appropriate steps to take for women of childbearing age diagnosed with sickle cell disease.

4.3.1 Age

Approximately 62.7% of the participants were ages 18 to 20, 62% were ages 21 to 25, and 36% were ages 26 to 29. Generally, all age groups showed prior knowledge of sickle cell disease. The number of correct responses decreased when students were asked
specific questions regarding the incidence and prevalence rate of sickle cell disease, as shown in Table 11. Only 15.4% of the students ages 18 to 20, 6.2% of the students ages 21 to 25, and 33.3% of students ages 26 to 29 correctly answered Question 3, which asked: “In the United States, how many babies are born with sickle cell disease per year?” Only 37.6% of the students ages 18 to 20, 43% of the students ages 21 to 25, and 66.6% of the students ages 26 to 29 correctly responded to Question 9, which asked: “What is the estimated number of people living with sickle cell disease in the United States?” No statistical differences in the variation of response rates occurred between the three age groups.

4.3.2 Gender

A total of 30.2% males and 69.3% females completed the sickle cell disease questionnaire. Both genders had a general understanding of sickle cell disease from a medical standpoint. However, results showed that 20.5% of males and 15.7% of females were able to identify how many babies were born in the United States per year with sickle cell disease. The results showed that 38.5% of males and 40.7% of females responded correctly to the number of people living in the United States with sickle cell disease.

In addition, only 48.7% of male students and 42.5% of female students were able to correctly identify the estimated life expectancy of people living with sickle cell disease. Question 4 was significant for the Pearson’s chi-square result (p = .047). A significant difference occurred in knowledge between males and females when compared to previously discussed questions. As shown in Table 10, male and female students were
more likely to respond correctly to the following questions: Question 10: “Is there a cure for sickle cell disease”; Question 11: “What population is more at risk for developing sickle cell disease”; Question 12: “Sickle cell anemia is a medical condition where the red blood cells ____?; and Question 13: “When is the appropriate time to test for sickle cell disease?” No statistical differences in the variation of response rates occurred between male and females for questions 11 to 14.

4.3.3 Ethnicity

Table 9 also illustrates that 85.1% of Caucasian students, 78.9% of African-American students, 75% of Hispanic students, and 71.1% of “Other” students were knowledgeable about the origins of sickle cell disease. Interestingly, 18.4% of Caucasian students, 7% African-American students, 18.7% of Hispanic students, and 24.4% of “Other” students correctly responded to Question 3: “In the United States, how many babies are born with sickle cell disease per year?” Only 41.8% of Caucasian students, 47.4% of African-Americans students, 37.5% of Hispanic students and 48.8% of “Other” students were aware of the average life expectancy of people living with sickle cell disease (Question 4).

The percentages of Caucasian students who correctly identified the medical complications caused by sickle cell disease were 85.8%, 84.2 % for African-Americans, 87.5% for Hispanic students, and 84% for “Other” students. Additionally, 38.6% of Caucasian students, 47.4% of African-American students, 37.2% of Hispanic students, and 38.6% of “Other” students responded correctly to the number of people living in the
United States with sickle cell disease (Question 9). Approximately 95% of Caucasian
students, 96.5% of African-American students, 100% of Hispanics, and 88.8% of “Other”
students identified that no cure for sickle cell disease exists (Question 10). In addition,
42.1% of Caucasians students, 40.1% of African-American students, 56.2% of Hispanic
students, and 48.8% of “Other” students could correctly identify the appropriate actions
for women of childbearing age, who are diagnosed with sickle cell disease, to take when
trying to conceive. This study showed that no statistical difference existed in the variation
of response rates between ethnicities.
CHAPTER 5
DISCUSSION

5.1 Study Findings

The research study was designed to examine whether or not college students attending the University of Illinois at Urbana-Champaign (UIUC) had any prior knowledge of sickle cell disease. Three research questions were used as the primary framework for this study. Research Question 1 asked, “How knowledgeable are midwestern college students on background information regarding sickle cell disease?” Study results revealed that participants had some existing knowledge of this disease. However, among the sample group of college students, all of them scored the lowest when questioned on the prevalence and incidence rates of this disease (see Table 6). Only 17% of the students answered Question 3 correctly, which asked, “In the United States, how many babies are born with sickle cell disease per year?” Thirty-nine percent of the students responded correctly to Question 9, which asked, “What is the estimated number of people living with sickle cell disease in the United States?”

Moreover, findings from this study are similar to other published sickle cell disease studies concerning participants being more aware of the patterns of inheritance of SCD, as well as the prevalence and incidence rates of SCD. A research study conducted by Boyd et al. (2005) showed that students scored low on questions that focused on the inheritance patterns of SCD. Boyd et al. (2005) showed that 9% (15/162) of the participants understood the inheritance pattern of SCD. In addition, in a study by Dyson
(1997), only 25% of the participants correctly answered questions on the inheritance patterns of SCD. These results indicate that individuals are unaware that they could be carriers of this disease and could be at risk of producing an offspring with SCD or the sickle cell trait. The basic knowledge of knowing how many people are born and living with this disease is crucial. Again, Table 6 (see Appendix B) shows that college students are not familiar with how many people are living with this disease. The results from this study showed that only 17% of UIUC students correctly identified how many babies are born with sickle cell disease per year, and 39.8% correctly responded to the number of people living with sickle cell disease in the United States. It is imperative for people to be aware of their SCD or SCT status, as well as knowing general information about this disease.

Research Question 2 in the study asked, “Do any significant differences exist in awareness between ethnic groups or groups of students who have a higher probability of inheriting sickle cell disease traits?” Results showed no significant differences between ethnic groups, especially among African-American students who are most at risk of inheriting the sickle disease trait. In this study, African-American students failed to know more about sickle cell disease than their Caucasian, Hispanic, and “Other” classmates. Therefore, this research study illustrates that college students have some existing knowledge about sickle cell disease. Yet the overall reviewed literature on sickle cell disease collectively stresses that a dire need remains for more extensive research and awareness programs to reach at-risk populations on the severity of this disease and the importance of knowing if an individual is a carrier of the sickle cell trait. It is imperative
that at-risk populations are well informed on the medical complications caused by sickle cells disease, as well as their carrier status. This information will help them make an informed decision on reproduction and genetic counseling options.

Research Question 3 asked, “Do any significant differences exist in awareness between gender groups?” Results showed a significant difference in response rates for males and females who answered Question 4, which asked, “What is the estimated life expectancy of people living with sickle cell disease?” The statistical difference for Question 4 was (p = .047), and therefore only 42.5% (see Appendix B) of the females answered the question correctly while 48.7% of the males responded correctly.

By and large, the estimated life expectancy for people living with SCD is the mid-40s and 50s while the average life expectancy for humans without SCD ranges from the early to mid-70s. People living with sickle cell disease are estimated to lose 30 years of life in comparison to others living without this disorder. The estimated difference is great, and it is crucial for the public to know the biological and physical effects of sickle cell disease. In addition, people who develop this disease suffer from medical complications and costly medical bills. They are constantly seeking medical treatments in order to live a normal and productive life. To have a shortened life span due to this disease can be tragic. Continued research on sickle cell disease must be done to help improve the lives of people stricken with this disorder. It is also important for our generation to know how short the life expectancy is for people living with sickle cell disease, as well as the severe health problems sickle cell individuals face throughout their lifetime.
5.2 Limitations

As with any research study, this particular study had several limitations. This research study was designed only to sample students enrolled in three Community Health courses. Surveying students in Community Health courses limited the participant sample and excluded other university students who were less knowledgeable about this disease because they were not exposed to this medical disease in classes and/or at a risk of being a carrier of the sickle cell disease trait. Different results may have been obtained in a larger sample of students who were not all enrolled in Community Health classes.

This research study’s questionnaire also had several limitations. The questionnaire failed to include questions that would have specifically inquired how and when participants became knowledgeable about SCD. These questions are important and could be used to help determine the preferred method to educate college students about SCD. Thus, such information could provide a framework for developing additional health education programs on sickle cell disease on campus and in the community. Not only was it important to question how students became knowledgeable about this disease, but also if they had the disease or knew anyone with the disease. That valid information would have helped determine if they were familiar with the disease due to being exposed to a person living with SCD.

The questionnaire failed to capture the marital status and or sexual relationship of participants. This information would have allowed further analysis of at-risk populations’ awareness of SCD. This information would have also indicated a possible need for more
research on campus on SCD genetic and counseling for the university students. In addition, these results reflected responses only from college participants’ ages of 18 to 30. The awareness in this segment is important because the majority of births occur within this age range, and it is important for all young adults to be aware of this disease. If this study were extended to all participants in this age range, it would have more validity in the response rate. The questionnaire also did not record the participant’s economic background or status. Most people with a higher socioeconomic background are most likely to seek a physician’s care for health and medical issues due to their economic means. People from a lower socioeconomic background may not have the resources to follow up with a physician and seek proper care due to their lack of finances. Lastly, the questionnaire did not include a segment on SCD inheritance patterns. The inheritance patterns are important when producing offspring. Couples must be aware that the probability is higher for producing a child with sickle cell disease when both are carriers for the disease.

5.3 Suggestions

More research on the public’s general knowledge about this disease will help determine the areas where more education is needed on SCD. The reviewed literature has focused on the biological genetics of this disease, but not much research has specifically addressed the awareness of this disease in all populations. Further studies are needed to address the effectiveness of intervening media campaigns to increase sickle cell awareness. It is important to evaluate which media source is most effective in increasing
the public’s awareness on this disease. We live in a technological age where Facebook, Twitter, MySpace, television/public radio commercials, web advertisement, and celebrity spokesmen are all currently a part of the marketing strategies used to inform the general public about a health disease or product. If this same approach was used to increase public awareness, more people would be informed about SCD, as well as their status as a carrier.

Another factor that can help address this issue is placing a public figure in the forefront for sickle cell disease. Ideally, he or she could help start a movement on SCD awareness. The public figure would more than likely generate additional funds for research, treatment, and even cures for SCD. Public interest would increase more medical training on this particular disease and help eliminate attitudes and preconceived notions of this disease. With more education and training, patients will be able to receive optimum care, and more centers can be built to specifically treat this disease in high-risk communities.

More research on health care professionals’ perceptions and practices on pain-related SCD would also help increase sickle cell disease awareness in the medical world. The reviewed literature shows that some pain specialists who operate on sickle cell patients have inadequate knowledge on the disease’s related pain. Therefore, physicians must continue to build strong relationships with patients who have SCD. Establishing centers that focus on this disease will allow more people to receive appropriate treatment and care.
In addition, studies have not addressed sickle cell disease and interracial marriages. In the United States, we see a steady growth of immigrants from all over the world. Our world has become more diverse, and more couples are pushing the racial boundaries and marrying outside their race (see Figure 2-7). Researchers from the Pew Research Center for the People and the Press, an independent research organization, reported that one out of seven new marriages are multi-racial or multi-ethnic (Chen, 2010).

![Growing diversity](image)

Figure 5-1: Interracial marriages 2005 (Microsoft News, 2007).

An increase in interracial marriages among the millennial generation 18 to 29 year olds has occurred. Research shows that 85% of the millennial generation accepts interracial marriages, and more people from this age group are producing a new
generation of multi-racial children (Chen, 2010). Since interracial marriages are a growing trend, a need arises for more research on interracial marriages and how they correlate with sickle cell disease. Sickle cell disease will soon have an impact on all ethnicities and the gene will no longer be found primarily among African-Americans. Since this disorder will soon be a problem for all races, it is important that all ethnicities are knowledgeable about this disease and the effects that SCD has on the human body.

In contrast to Chen (2010), Bloom (1995) argued that, due to natural selection, the Hb S gene has become less common among African-Americans in comparison to their African ancestors. Bloom (1995) suggested that fewer African-Americans will have this particular trait. He believed that interracial mating and the decrease of needing a survival mechanism against malaria are causing the gene to be diluted over time. Bloom (1995) believed that these two factors explain why a reduction in the frequency of SCD in black Americans will occur.

On the other hand, with an increase of sickle cell disease in our society, health care will became a main issue. An individual with this disease suffers from chronic health problems throughout his/her life. It is therefore imperative for treatment to be accessible to the public regardless of race, religion, gender, and sexual orientation. With a possible influx of urgent treatment for people with sickle cell disease, more SCD medical centers will need to be established. With sickle cell disease awareness in our communities and federal funding for addition SCD research, plus our government intervention to put a plan
into action to cover SCD medical expenses, will help the process of finding a cure to eliminate SCD around the world.
References


APPENDIX
Reference Tables
Table 1: Incidence of Sickle Cell Disease Genotypes

<table>
<thead>
<tr>
<th>Genotypes</th>
<th>% of SCD Children</th>
<th>Clinical Severity</th>
<th>Type of Hemoglobin %</th>
</tr>
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<tbody>
<tr>
<td>Hb SS</td>
<td>71.9%</td>
<td>Usually Marked</td>
<td>S 80-90</td>
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<td>Hb S HPF</td>
<td>0.1%</td>
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<td>Hb SC</td>
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<td>Mild to moderate</td>
<td>S 50-90</td>
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<td>Hb SA</td>
<td>Asymptomatic</td>
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Table 2: Cost-Effectiveness of Hydroxyurea

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<th>Item</th>
<th>Cost</th>
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<tr>
<td>Mean Hospitalization Cost for Painful Crises without Hydroxyurea Regimen</td>
<td>$17,290</td>
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<tr>
<td>Mean Hospitalization Cost for Painful Crises with Hydroxyurea Regimen</td>
<td>$12,160</td>
</tr>
<tr>
<td>Annual Average Hospital Cost per Patient on hydroxyurea</td>
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<td>Annual Average Hospital Cost per patient not on Hydroxyurea</td>
<td>$22,020</td>
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APPENDIX B
Study Results Tables
Table 3: Respondents to Sickle Cell Questionnaire

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<th>Age of respondents</th>
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<table>
<thead>
<tr>
<th>Ethnic origin of respondents</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>African-American</td>
<td>57</td>
<td>22.0</td>
</tr>
<tr>
<td>Caucasian</td>
<td>140</td>
<td>54.0</td>
</tr>
<tr>
<td>Hispanic</td>
<td>16</td>
<td>6.0</td>
</tr>
<tr>
<td>Other</td>
<td>45</td>
<td>17.0</td>
</tr>
</tbody>
</table>

Note: For “Other” students, students, were able to identify themselves by filling in the blank for ethnicity.

Table 4: Percentage of Correct Responses for Biological Aspect of Sickle Cell Disease

<table>
<thead>
<tr>
<th>Question #</th>
<th>Question</th>
<th>Percent (N)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>What is sickle cell disease?</td>
<td>92.7 (259)</td>
</tr>
</tbody>
</table>
### Table 5: Percentage of Correct Responses for Sickle Cell Disease Origin

<table>
<thead>
<tr>
<th>Question #</th>
<th>Question</th>
<th>Percent (N)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>Sickle cell disease evolved in which of the following continents?</td>
<td>80.7 (258)</td>
</tr>
<tr>
<td>12</td>
<td>Sickle cell anemia is a medical condition where the red blood cells…?</td>
<td>78.8 (258)</td>
</tr>
</tbody>
</table>

### Table 6: Percentage of Correct Responses for Incidence and Prevalence Rates of Sickle Cell Disease

<table>
<thead>
<tr>
<th>Question #</th>
<th>Question</th>
<th>Percent (N)</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>In the United States, how many babies are born with sickle cell disease per year?</td>
<td>17 (258)</td>
</tr>
<tr>
<td>4</td>
<td>What is the estimated life expectancy of people living with sickle cell disease?</td>
<td>44 (259)</td>
</tr>
<tr>
<td>9</td>
<td>What is the estimated number of people living with sickle cell disease in the United States?</td>
<td>39.8 (257)</td>
</tr>
</tbody>
</table>
Table 7: Percentage of Correct Responses for Medical Aspects of Sickle Cell Disease

<table>
<thead>
<tr>
<th>Question #</th>
<th>Question</th>
<th>Percent (N)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5</td>
<td>What medical complications are caused by sickle cell disease</td>
<td>84.9 (257)</td>
</tr>
<tr>
<td>10</td>
<td>Is there a cure for sickle cell disease?</td>
<td>94.2 (258)</td>
</tr>
<tr>
<td>11</td>
<td>What population is more at risk for developing sickle cell disease?</td>
<td>91.1 (257)</td>
</tr>
</tbody>
</table>

Table 8: Percentage of Correct Responses for Genetic Counseling and Screening

<table>
<thead>
<tr>
<th>Question #</th>
<th>Question</th>
<th>Percent (N)</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>When is the appropriate time to test for sickle cell disease?</td>
<td>89.2 (258)</td>
</tr>
<tr>
<td>14</td>
<td>What is the appropriate action for a person of child-bearing age diagnosed with sickle cell disease?</td>
<td>43.6 (258)</td>
</tr>
</tbody>
</table>

Note: All participants are of childbearing age.
Table 9: Percentage of Correct Responses by Participants Grouped by Ethnicities

<table>
<thead>
<tr>
<th>Ethnicities</th>
<th>N</th>
<th>Q1</th>
<th>Q2</th>
<th>Q3</th>
<th>Q4</th>
<th>Q5</th>
<th>Q9</th>
<th>Q10</th>
<th>Q11</th>
<th>Q12</th>
<th>Q13</th>
<th>Q14</th>
</tr>
</thead>
<tbody>
<tr>
<td>African-American</td>
<td>57</td>
<td>96.5</td>
<td>78.9</td>
<td>7.0</td>
<td>47.4</td>
<td>84.2</td>
<td>47.4</td>
<td>96.5</td>
<td>93.0</td>
<td>80.7</td>
<td>87.7</td>
<td>40.4</td>
</tr>
<tr>
<td>Caucasian</td>
<td>140</td>
<td>93.6</td>
<td>85.1</td>
<td>18.4</td>
<td>41.8</td>
<td>85.8</td>
<td>8.6</td>
<td>95.0</td>
<td>92.8</td>
<td>81.4</td>
<td>90.7</td>
<td>42.1</td>
</tr>
<tr>
<td>Others</td>
<td>45</td>
<td>95.9</td>
<td>71.1</td>
<td>24.4</td>
<td>48.8</td>
<td>84.0</td>
<td>38.6</td>
<td>88.8</td>
<td>86.6</td>
<td>77.7</td>
<td>93.3</td>
<td>48.8</td>
</tr>
<tr>
<td>Hispanic</td>
<td>16</td>
<td>94.7</td>
<td>75.0</td>
<td>18.7</td>
<td>37.5</td>
<td>87.5</td>
<td>31.2</td>
<td>100</td>
<td>93.7</td>
<td>56.2</td>
<td>75.0</td>
<td>56.2</td>
</tr>
</tbody>
</table>

Table 10: Percentage of Correct Responses by Correct Answers Grouped by Age

<table>
<thead>
<tr>
<th>Age</th>
<th>N</th>
<th>Q1</th>
<th>Q2</th>
<th>Q3</th>
<th>Q4</th>
<th>Q5</th>
<th>Q9</th>
<th>Q10</th>
<th>Q11</th>
<th>Q12</th>
<th>Q13</th>
<th>Q14</th>
</tr>
</thead>
<tbody>
<tr>
<td>18-20</td>
<td>162</td>
<td>93.2</td>
<td>79.6</td>
<td>15.4</td>
<td>43.8</td>
<td>85.1</td>
<td>37.6</td>
<td>95.6</td>
<td>90.7</td>
<td>79.0</td>
<td>88.8</td>
<td>44.4</td>
</tr>
<tr>
<td>21-25</td>
<td>93</td>
<td>91.3</td>
<td>82.7</td>
<td>6.2</td>
<td>45.1</td>
<td>86.0</td>
<td>43.0</td>
<td>92.4</td>
<td>91.3</td>
<td>79.5</td>
<td>51.2</td>
<td>40.8</td>
</tr>
<tr>
<td>26-29</td>
<td>3</td>
<td>100</td>
<td>100</td>
<td>33.3</td>
<td>33.3</td>
<td>66.6</td>
<td>66.6</td>
<td>66.6</td>
<td>100</td>
<td>100</td>
<td>100</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 11: Percentage of Correct Responses Answers Distributed by Gender

<table>
<thead>
<tr>
<th>Gender</th>
<th>N</th>
<th>Q1</th>
<th>Q2</th>
<th>Q3</th>
<th>Q4¹</th>
<th>Q5</th>
<th>Q9</th>
<th>Q10</th>
<th>Q11</th>
<th>Q12</th>
<th>Q13</th>
<th>Q14</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>78</td>
<td>93.6</td>
<td>74.4</td>
<td>20.5</td>
<td>48.7</td>
<td>81.8</td>
<td>38.5</td>
<td>91.0</td>
<td>91.0</td>
<td>73.1</td>
<td>92.3</td>
<td>38.5</td>
</tr>
<tr>
<td>Female</td>
<td>179</td>
<td>92.7</td>
<td>84.3</td>
<td>15.7</td>
<td>42.5</td>
<td>87.6</td>
<td>40.7</td>
<td>96.1</td>
<td>92.1</td>
<td>81.5</td>
<td>88.2</td>
<td>46.1</td>
</tr>
</tbody>
</table>

¹X² (3, N=259), 7.97, p=.047
APPENDIX C
IRB Approval Form
October 22, 2008

Susan Farmer  
Kinesiology & Community Health  
Rm 212a Huff Hall  
MC 548

RE: Sickle Cell Disease Awareness amongst College Students  
IRB Protocol Number: 09204

Dear Susan:

Thank you for submitting the completed IRB Application for Exemption form for your project entitled Sickle Cell Disease Awareness amongst College Students. Your project was assigned Institutional Review Board (IRB) Protocol Number 09204 and reviewed. The research activities involving human subjects are exempt from Title 45 - Public Welfare, Part 46 - Protection of Human Subjects, Subpart A - Federal Policy for the Protection of Human Subjects per the following category:

45 CFR 46.101(b)(2): This exemption applies since the study involves UIUC students enrolled in Community Health 260, 210, and 230 completing a questionnaire containing questions regarding sickle cell disease and demographic information. The goal of the study is to examine whether college students from a Midwestern Big Ten school are aware of sickle cell disease and can identify common symptoms, prevention methods, and risks of the disease. No information that could lead to individual identification is collected, therefore the category 2 exemption may be applied.

This determination of exemption only applies to the research study as submitted. Exempt protocols are approved for a maximum of three years. Please note that additional modifications to your project need to be submitted to the IRB for review and exemption determination or approval before the modifications are initiated. To submit modifications to your protocol, please complete the IRB Research Amendment Form (see http://www.irb.uiuc.edu/?p=3-forms-and-instructions/research-amendments.html).

We appreciate your conscientious adherence to the requirements of human subject research. If you have any questions about the IRB process, or if you need assistance at any time, please feel free to contact me or the IRB Office.

Sincerely,

[Signature]

Sat Keen, Director, Institutional Review Board

c: Cassie Jessica Osbourne

telephone 217-333-2670 • fax 217-333-0415 • email IRB@uiuc.edu
APPENDIX D
Informed Consent for Participants of the Questionnaire
Sickle Cell Disease

You are invited to participate in research being conducted by Susan Farner, PhD in the Department of Kinesiology and Community Health Department at the University of Illinois at Urbana-Champaign. The purpose of the research is to evaluate your awareness and knowledge of sickle cell disease. This information will be used to help determine how best to present information about this disease to college students. Your identification will not be used. The data will be aggregated without identification.

You are asked to complete a survey. You must be 18 years of age or older to participate in this research. The survey will take approximately 10 minutes to complete. You may skip any questions you are not comfortable answering. Your participation in the research is completely voluntary. Your decision to participate, decline, or withdraw from participation will have no effect on your status at the University of Illinois. The results of this research will be reported in Master Thesis projects, in journal articles and conference presentations.

The risks associated with this project not more than those of every day life. The benefits to this project will be to determine how best to present information about this disease to college students. You will be given a copy of the consent form for your records.

If at any time, you have questions about this research project, please feel free to contact the principal investigator, Susan Farner, PhD, Department of Kinesiology and Community Health, 129 Huff Hall, 333-6876, sfarner@uiuc.edu. You are welcome to call collect if you identify yourself as a research participant.

If you have any questions about your rights as a participant in research involving human subjects, please feel free to contact the University of Illinois Institutional Review Board (IRB) office at 217.333.2670 or irb@illinois.edu. You are welcome to call collect if you identify yourself as a research participant.

I am 18 years of age or older.
I have read and understand the above consent form and voluntarily agree to participate in this study.
APPENDIX E
Questionnaire
Sickle Cell Disease

Age __________

Gender  Male  Female

Ethnicity  Caucasian  African American  Hispanic  Other __________

Please select the one best answer.

1.) What is sickle cell disease?
   A. Inherited blood disorder
   B. Infectious disease
   C. Sexually transmitted disease
   D. Fatal illness

2.) Sickle cell disease evolved in which of the following continents?
   A. Africa
   B. Asia
   C. North America
   D. South America

3.) In the United States, how many babies are born with sickle disease per year?
   A. 1,000
   B. 200,000
   C. 7,000,000,000
   D. 500,000

4.) What is the estimated life expectancy of people living with sickle cell disease?
   A. 40 years
   B. 75 years
   C. 35 years
   D. 20 years
5.) What medical complications are caused by sickle cell disease?
   A. Stroke
   B. Lung tissue damage
   C. Pain episode
   D. All of the above

9.) What is the estimated number of people living with sickle cell disease in the United States?
   A. 2.5 million
   B. 1 million
   C. 4 million
   D. 6 million

10.) Is there a cure for sickle cell disease?
     A. Yes
     B. No

11.) What population is more at risk for developing sickle cell disease?
     A. Caribbean Island
     B. African
     C. Saudi Arabian
     D. Mediterranean

12.) Sickle cell anemia is a medical condition where the red blood cells ....
     A. have an unusual shape
     B. have a higher than normal number
     C. are no longer produced by the body
     D. none of the above
13) When is the appropriate time to test for sickle cell disease?
   
   A. Newborn screening programs
   B. Doctor check ups
   C. At HIV testing
   D. At Sexually transmitted disease testing

14. What is the appropriate action for a person for child bearing age diagnosed with sickle cell disease?

   A. Preventive medicine
   B. Exercise
   C. Genetic counseling
   D. Vitamins