

# KNOWLEDGE OF SICKLE CELL TRAIT AND DISEASE AMONG AFRICAN-AMERICAN COLLEGE STUDENTS

by

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The term sickle cell disease (SCD) refers to a collection of hemoglobinopathies (inherited blood disorders) characterized by abnormal hemoglobin and produced through the homozygous inheritance of a sickle cell allele. Heterozygous inheritance results in a condition known as sickle cell trait (SCT). Individuals with SCT have a 50% chance of passing the gene to future offspring. In recent years mounting evidence has confirmed that SCT is also associated with several rare but serious complications including renal complications, renal cancer, spleen damage, and exercise-related sudden death. In the United States, sickle cell conditions primarily occur among African Americans. While all 50 states conduct newborn screenings which identify individuals with SCD and SCT, no studies have examined whether trait status is effectively conveyed to affected individuals or investigated knowledge of SCT among a large sample of African Americans. The purpose of the present study was to examine knowledge of SCT and SCD and to identify whether current procedures for trait notification in North Carolina effectively convey information about trait status, as well as its health and reproductive implications. A large sample of African-American college students ( $N = 258$ ) completed questionnaires assessing knowledge of SCT and SCD. Participants reported their trait and disease status, the status of family members, and sources of sickle cell knowledge. Results indicated that participants were most likely to have received information about sickle cell from

school. Though participants were generally familiar with the terms “sickle cell disease” and “sickle cell trait,” many lacked knowledge regarding the genetic transmission of SCD, common symptoms, and treatment. A majority of participants were uncertain of their SCT status. Unfortunately, reported trait status of the participants could not be verified due to missing records. Nonetheless, participants who indicated that they had SCT or “thought” they had SCT scored higher on a measure of trait knowledge. Participants who had received information about sickle cell from their families showed greater trait knowledge than those who had not. Females were more likely than males to desire to know their trait status. Females also displayed higher levels of trait and disease knowledge than males.



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## Chapter I: Introduction

### Sickle Cell Disease

The term sickle cell disease (SCD) refers to a collection of congenital hemoglobinopathies (i.e., inherited blood disorders) which are characterized by abnormal hemoglobin (Hb) molecules (National Institutes of Health, 2002). Hemoglobin is the protein within erythrocytes (i.e., red blood cells), which is responsible for collecting oxygen in the lungs and transporting it throughout the body. A molecule of hemoglobin typically consists of four protein subunits: two subunits of beta-globins (i.e.,  $\beta$ -globin) and two subunits of alpha-globins (i.e.,  $\alpha$ -globins). Each of these four protein subunits carries an iron-containing molecule called heme, which is necessary for the red blood cell to collect oxygen in the lungs and transport it throughout the body. Individuals typically inherit two genes which code for normal beta-globin, one from the mother and one from the father. Rarely, individuals inherit two abnormal, mutated beta-globin sickle genes, resulting in a condition known as sickle cell disease (SCD).

In an individual with SCD, the abnormal beta-globin molecules impair the blood's ability to efficiently transport oxygen to the rest of the body and, under certain conditions, can cause sickling of the erythrocytes (National Institutes of Health, 1996). Normally, erythrocytes have a smooth, rounded shape and can easily bend and flex in order to glide through the body's capillaries, carrying oxygen throughout the body. However, for individuals with SCD, the abnormal beta-globin causes erythrocytes to become malformed, taking on a half-moon, sickle-like appearance. Most commonly, rather than coding for the normal beta-globin subunit, the mutated hemoglobin produces an abnormal version of beta-globin, called hemoglobin S (i.e., HbS). Inheriting two genes which code for HbS results in the most prevalent form of SCD (i.e., HbSS). Other mutations in the beta-globin gene can result in less-common forms of sickle cell disease (i.e., HbSC, HbS beta-thalassemia).

In contrast to normal, healthy erythrocytes, “sickled” red blood cells are rigid and sticky and cannot travel easily through tiny blood vessels. In fact, the sickle-shaped erythrocytes can block the blood vessels and restrict the flow of blood through the body, resulting in a condition known as vaso-occlusion. Vaso-occlusion can occur anywhere in the vascular system and may restrict or entirely cut off the blood supply necessary for healthy functioning of organs. When vaso-occlusion occurs, tissues in the affected area become deoxygenated, resulting in intense, unpredictable pain in body organs or joints, a condition termed “pain episodes” or “pain crises.” The frequency and severity of pain episodes can vary significantly among patients ranging from mild, short-lasting pain which may be treated with over-the-counter analgesics to severe, chronic pain requiring hospitalization and the administration of intravenous painkillers.

In addition to causing vaso-occlusion, sickled erythrocytes have a very short lifespan. While normal red blood cells typically remain viable in the bloodstream for around 120 days, sickled cells last for only 10 to 20 days. Because the body cannot replace them at a fast enough rate, an individual with SCD may experience chronic anemia (National Institutes of Health, 1996).

In the United States, the prevalence of sickle cell disease is estimated to be around 72,000 cases, with the disease occurring in approximately 1 in every 500 African-American births and 1 in every 1,000 to 1,400 Hispanic births (National Institutes of Health, 1996). Worldwide, the World Health Organization (2006) estimates that about 300,000 infants are born with major hemoglobinopathies each year, with SCD being the most prevalent. Because individuals with SCD display reduced incidence of malarial infection, the disease is more prevalent among individuals whose ancestry can be traced to regions in which malaria is or was endemic. Hence

SCD is particularly common among people whose ancestors lived in the regions of sub-Saharan Africa, India, Saudi Arabia, and the Mediterranean (World Health Organization, 2006).

Sickle cell disease is an autosomal recessive genetic disorder, thus an individual must inherit two recessive genes (i.e., one from each parent) which each code for abnormal beta-globin (HbS) in order for the disease state to occur. If a person receives a normal beta-globin gene (HbA) from one parent and a sickle beta-globin gene (HbS) from the other, that person is said to have sickle cell trait (SCT) or to be a “carrier” of the disease. Individuals with SCT are generally healthy, but have a 50% chance of passing the trait on to their children. As with all autosomal recessive genetic disorders, if each parent has one sickle beta-globin gene and one normal beta-globin gene (HbAS + HbAS), the couple would then have a 25% chance of producing a child with SCD (HbSS), a 25% chance of producing a child who does not have the disease or trait (HbAA), and a 50% chance of producing a child with SCT (HbAS). If one parent has SCD (HbSS) and the other parent has normal beta-globin (HbAA), then all offspring would have SCT (HbAS). Finally, if a parent has SCD (HbSS) and the other has trait (HbAS), each child would have a 50% chance of having SCT and a 50% chance of having SCD.

**Forms of Sickle Cell Disease.** There are several different types of SCD defined by which abnormal beta-globin gene has been inherited. In the United States, the three most prevalent forms of SCD are HbSS, HbSC, and HbS beta-thalassemia. The most common form of the inherited hemoglobinopathies responsible for SCD is homozygous SCD (HbSS) in which a person inherits two sickle cell genes (HbS), one from each parent. Other forms include HbSC, in which the individual inherits one sickle cell gene (HbS) and one gene for another type of abnormal hemoglobin (HbC), and HbS beta-thalassemia, in which the individual inherits one sickle cell gene (HbS) and one beta thalassemia gene. HbSC usually causes a milder form of the

disease, resulting in less frequent and less intense pain crises. Individuals with HbS beta thalassemia may be differentiated by whether the beta thalassemia gene codes for reduced beta globin production ( $\beta^+$ ) or no beta production ( $\beta^0$ ), with HbS  $\beta^0$  being the more severe form. Rarer forms of sickle cell disease also exist including HbSD, HbSE, and HbSO Arab, in which affected individuals inherit one sickle cell gene and one gene which codes for another abnormal type of hemoglobin (e.g., D, E, or O).

**Complications of Sickle Cell Disease.** As previously mentioned, the most common symptom of sickle cell is a pain episode, also known as a sickle cell pain crisis, which occurs when tissue becomes damaged as a result of vaso-occlusion, resulting in inflammation and extreme pain. Sickle cell pain episodes may occur anywhere in the body and vary greatly in severity, location, and duration among patients (Ballas, 2007). Pain episodes are the most frequent cause of hospitalization for individuals with SCD (Ballas & Lusardi, 2005).

While pain episodes often occur in a person's joints, blood vessels in the hands and feet may also become blocked, resulting in pain, swelling, and fever. This condition, hand-foot syndrome, is often the first noticeable symptom to occur in infants with SCD. Eye problems, including retinal deterioration and blindness, may also occur making it extremely important that children with SCD receive regular vision exams. Sickle cell disease can also cause a shortage of erythrocytes. The scarcity and reduced life span of the red blood cells can lead to symptoms of jaundice (e.g., yellowing of the eyes, yellowish tint to skin), as well as symptoms of anemia including fatigue, paleness, and shortness of breath. Children with SCD are also often small for age and experience delayed onset of puberty caused by the shortage of red blood cells.

Some of the more severe complications of SCD include stroke, infection, and acute chest syndrome. Twenty-four percent of patients with SCD suffer a stroke (interrupted blood flow to

the brain) by the age of 45, and because African-Americans have an already-heightened risk for stroke, those with SCD are particularly vulnerable (Verduzco & Nathan, 2009). In addition, the repeated episodes of ischemia (restricted blood flow) caused by sickled erythrocytes cause significant damage to the spleen, resulting in atrophy which is evident as early as six months of age (Khatib, Rabah, & Sarnaik, 2009). The spleen functions to filter old and damaged cells from the blood and also serves to produce antibodies. When it is impaired, the body is at increased risk of infection. Such infections may spread rapidly throughout the body, and without a properly functioning spleen, individuals with SCD are unable to fight and filter bacteria, leading to a high likelihood of sepsis (Booth, Inusa, & Obaro, 2010). Bacterial infections pose a risk for mortality and can also trigger severe pain episodes through a variety of pathophysiological processes (Booth, Inusa, & Obaro, 2010). While sepsis was once thought to be unavoidable, a landmark investigation of penicillin in the 1980s showed that prophylactic administration of penicillin decreases the rate of mortality from sepsis in young children with SCD (John et al., 1984). In addition, two emerging treatments—hydroxyurea, a chemotherapeutic agent, and stem cell transplantation—show promise in actually reversing splenic damage (Khatib, Rabah, & Sarnaik, 2009).

Infections can also result in acute chest syndrome, the leading cause of death in patients with SCD. Acute chest syndrome is a life-threatening complication which can occur when sickled erythrocytes become trapped in the lungs (pulmonary infarction) or as a result of a fat embolism or infection in the lungs (Vichinsky et al., 2000). Symptoms of acute chest syndrome may include generalized pain, chest pain, fever, and abnormal chest x-rays. Acute chest syndrome requires immediate hospitalizations and intubation is often needed during the early course of the illness.

**Prognosis.** Symptoms of sickle cell disease typically appear around five months of age, as the levels of fetal hemoglobin (the primary oxygen transporting protein present during fetal development) decline. Individuals with sickle cell have reduced life expectancy, with most early deaths occurring either from bacterial infection during infancy or from stroke or acute chest syndrome later in life (Davis, Schoendorf, Gergen, & Moore, 1997). Present life expectancy for individuals with SCD in the United States continues to be significantly reduced, with males having a median life expectancy of 42 years and females, 48 years (Platt et al., 1994).

### **Sickle Cell Trait (SCT)**

Individuals with SCT (one normal beta-globin gene and one abnormal sickle gene) generally experience few complications and lead healthy, full lives. However, they may pass the sickle cell gene to their own children; thus knowledge of trait status and education regarding transmission of the gene is critical if individuals with SCT wish to engage in reproductive planning. Over 300 million people worldwide are estimated to have SCT. While the highest prevalence of carriers is found in Africa and the Mediterranean region, SCT is also prevalent among individuals of African descent in the United States. Prevalence rates of SCT in the United States are estimated to be 8% among African-Americans compared to 0.05% among Caucasians (Tsaras, Owusu-Ansah, Owusua-Boateng, & Amoateng-Adjepong, 2009).

### **Complications of SCT**

Sickle cell trait is generally a benign medical condition, and most individuals with SCT are healthy and have a typical lifespan. As a result, a person with SCT may be unaware of their positive trait status. However, an individual with SCT has a 50% chance of passing on the abnormal sickle cell gene to any future offspring, making knowledge of trait status critical if the individual desires to engage in informed reproductive decision making.

In addition, there are some rare complications associated with SCT that make it critical for affected individuals and their primary care physicians (PCPs) to be aware of positive trait status. Compelling evidence began to accumulate in the 1940s and 1950s when several case studies were published suggesting a causal role for SCT in some rare sudden deaths, though at that time the distinction between SCT and SCD was not clearly understood. As early as 1940, the recommendation was made that all African-American patients should be tested for SCT, then referred to as “sickleemia,” before undergoing surgery due to the risk of erythrocytes sickling under hypoxic conditions (Bauer, 1940). This recommendation was made after several individuals with SCT died suddenly and unexpectedly during surgery. Similarly, Adams (1957) suggested an increased risk in sudden death during pregnancy among women with trait. As evidence mounted, it became increasingly clear that while SCT represents a benign condition for most affected individuals, it does pose increased risk for a variety of negative outcomes, particularly under certain conditions.

Tsaras et al. (2009) reviewed evidence in order to define definite, probable, possible, and unlikely medical complications which are associated with SCT. The authors asserted that while SCT is primarily a protective carrier state which offers resistance to malaria, it is associated with several negative health outcomes. Definite associations include renal medullary cancer, hematuria (blood in the urine), renal papillary necrosis, hypothesnuria (reduced ability to concentrate urine), splenic infarction, exertional rhabdomyolysis (explosive muscle breakdown), and exercise-related sudden death (Tsaras et al., 2009).

Renal medullary carcinoma is a rare and highly aggressive tumor of the kidney that occurs almost exclusively in young adults with SCT. In 1995, this cancer was first reported in a series of case studies. Davis, Mostofi, and Sesterhenn (1995) identified 34 incidents of the

cancer, 33 of which were found in individuals with SCT. Since that initial report, approximately 120 total cases have been identified, only one of which occurred in an individual who did not have SCT (Watanabe et al., 2007). Within affected individuals, a tumor forms at the distal collecting ducts of the kidney and quickly spreads into the renal sinuses. Patients initially may experience symptoms including hematuria and flank pain. The cancer progresses rapidly with median survival rate from time of diagnosis being 15 weeks (Tsaras et al., 2009).

Other kidney problems including hematuria, renal papillary necrosis, and hyposthenuria also occur with increased frequency among individuals with SCT, with hematuria being the most frequently observed complication of SCT (Kiryluk, Jadoon, Gupta, & Radhakrishnan, 2007). Hematuria (blood in the urine) is frequently due to renal papillary necrosis, a disorder of the kidney in which the tissues of the renal papillae die. Hematuria can also result from kidney infection, kidney stones, von Willebrand's disease, or a malignancy (Tsaras et al., 2007). Bleeding may range from mild to severe. The renal papillae are the areas where the collecting ducts enter the kidney. When this area becomes atrophied, the kidney is unable to concentrate urine, a symptom known as hyposthenuria, which is sometimes also experienced by individuals with SCT. Individuals experiencing hyposthenuria have reduced stores of water in the body. When they do not compensate for this by drinking more fluids, the blood's plasma may become too concentrated (osmolastic), one factor linked with exertional exercise-related sudden death (Tsaras et al., 2007).

In addition to kidney complications, individuals with SCT are at increased risk for splenic infarctions, when the vessels of the spleen become congested by sickled erythrocytes (Tsaras et al., 2007). While usually mild, severe splenic infarction is more likely to occur at high altitudes, when an individual is exposed to low oxygen levels. Symptoms of splenic infarction include

severe abdominal pain, splenomegaly, and tenderness. Most mild cases of splenic infarction are treated with bed rest, analgesia, hydration, and occasionally oxygen. However, in severe cases, a carrier individual's spleen may rupture causing significant intraperitoneal bleeding, in which case a splenectomy is typically performed.

The most serious potential complication of sickle cell trait is exertional collapse and sudden death (Kerle & Nishimura, 1996). Though rare, sudden death may occur for individuals with SCT under certain conditions including severe hypoxia, acidosis, dehydration, and hypothermia. Under such extreme conditions, the erythrocytes of an individual with SCT can quickly sickle, increasing the chance for polymerization (bonding). Polymerization of the red blood cells then blocks blood vessels reducing the supply of oxygen to the body's muscles, which causes ischemic rhabdomyolysis or rapid muscle breakdown. Rhabdomyolysis causes serious metabolic problems and can result in sudden death.

Risk of this polymerization varies depending upon the level of hemoglobin S in the blood. Individuals with higher concentrations of HbS within the blood have an increased risk for polymerization. Therefore individuals with SCT are at increased risk for polymerization compared to those with typical hemoglobin, but are at reduced risk compared to individuals with SCD. Specifically, Lange, Minnich, and Moore (as cited in Tsaras et al., 2009, p. 508) demonstrated through in vitro studies that the erythrocytes of individuals with SCT become sickled when the blood's oxygen level is decreased to 2%, while the erythrocytes of individuals with SCD sickle when oxygen is reduced to 4-6%.

The sickling and polymerization of red blood cells puts individuals with SCT at risk for collapse when engaged in strenuous activities, such as vigorous physical exercise, because such activities reduce the level of oxygen within the body. Lack of oxygen triggers intra-vascular

sickling of the erythrocytes, which in turn causes vaso-occlusion (i.e., blood vessel blockage), resulting in further organ and tissue damage. This chain of events may quickly culminate in rhabdomyolysis, myocardial ischemia, arrhythmia, and sudden death (Scheinin & Wetli, 2009).

Sudden death resulting from SCT was originally described through several case studies of military recruits who collapsed during intense physical activity. Kark, Posey, Schumacher, and Ruehle (1987) reviewed exercise-related deaths and found a significant difference ( $p < .001$ ) for sudden death in individuals with SCT compared to those without the condition. Authors reviewed data available for more than 2 million enlisted recruits and found that the risk for sudden death during exercise increased by 28-fold for individuals with SCT compared to those without trait.

Numerous case studies have also been reported in which sudden death from SCT occurred in criminal suspects or incarcerated individuals following struggles and/or chases with police (Scheinin & Wetli, 2009). Suspects were typically engaged in chases or physical altercations with police when they suddenly collapsed; subsequent autopsies revealed massive intravascular sickling attributed to extreme physical activity.

Extreme physical activity can also pose a hazard for athletes with SCD. Although rare, athletes with SCT are at increased risk for exercise-related death (Tsaras, 2009). The National Athletic Trainer's Association (NATA) states that four physical states result in an increased risk of athletes with SCT experiencing the sickling of red blood cells: 1) severe hypoxemia, 2) metabolic acidosis, 3) hyperthermia in muscles, and 4) red-cell dehydration (NATA, 2007). The first known sudden death of an athlete with SCT occurred in 1974, when a college football player from Florida collapsed during a strenuous conditioning exercise at a high-altitude training camp in Colorado. The player collapsed after running sprints and died the next day; the cause of

death was determined to be acute exertional rhabdomyolysis associated with SCT (NATA, 2007). In fact, 13 college football players have died after a sickling collapse. NATA reports that nearly all of the 13 deaths in college football have been at institutions that did not screen for SCT or had a lapse in their prevention program (NATA, 2007). Collapses have typically occurred when athletes are “pushing their limits” and are involved in activities including sprints, running of stadium steps, intense sustained strength training, and running “gassers” or “suicides” in which players must complete a series of sprints without taking time to recover and breathe in between (NATA, 2007).

In 2006, Dale Lloyd, II, a football player at Rice University who had SCT collapsed and died following football practice as a result of sickling exertion. A lawsuit ensued between the Lloyd family and the National Collegiate Athletic Association (NCAA). The suit was settled in 2009 when the NCAA agreed to a series of actions to safeguard college athletes from sickle cell collapse. In June 2009, the NCAA agreed to recommend that all college and university athletic departments confirm SCT status in all student-athletes, if it is unknown, during their annual medical exams. This recommendation was made because, as the NCAA points out, although SCT screening is normally performed at birth, some student-athletes may not know their own status. In addition, the NCAA agreed to donate money to sickle cell research and to create an educational video about SCT (“The Student-Athlete with Sickle Cell Trait,” 2010).which was placed on the NCAA website (<http://www.ncaa.org>) along with several resources for student-athletes with SCT.

The NCAA recommendation aligns with a consensus statement released by the National Athletic Trainers Association (NATA) in 2007. A review of non-traumatic sports deaths in 1995 indicated that the top four causes of sudden death in high school and college athletes are (1)

cardiovascular conditions, (2) hyperthermia (heatstroke), (3) acute rhabdomyolysis associated with SCT, and (4) asthma (Van Camp, Bloor, Mueller, Cantu, & Olson, 1995). According to the NATA, from 2000 to 2007 at least nine high school or college athletes collapsed and died from exertional sickling, making it imperative that school and college administrators, coaches, and trainers are aware of the potential risks of strenuous exercise for young people with SCT in order to implement precautions for athletes with SCT (e.g., alternative workouts; NATA, 2007). Coaches and trainers should also be ready to respond appropriately and timely in case of a sickling emergency.

In addition to exertional sickling, there is also preliminary evidence that African-American women who use hormonal contraception may be at increased risk for venous thromboembolism (blood clot in a vein) if they possess SCT, although confirmatory studies are needed to verify this link (Austin et al., 2009). While hormonal contraception may put women with SCT at increased risk of negative outcomes, a PUBMED search for “sickle cell trait” and “death”, as well as “sickle cell trait” and “mortality,” revealed that more than 75% of case studies detailing SCT-related death have occurred in African-American adolescent boys and men, indicating that African-American males should be the prime target for trait awareness efforts (Ajayi, 2005).

### **History of Universal Newborn Screening for SCD**

Although health implications typically do not surface for SCT until adolescence or adulthood, the condition is usually identified as a result of newborn screening. The term newborn or neonatal screening is used to collectively refer to biochemical tests designed to identify the presence of inherited disorders, which are typically metabolic in origin (Therrell, 2001). Such tests occur during the first hours or days of an infant’s life and are used to detect a

variety of health problems. Newborn screenings began in the 1960s when the development of a blood screening procedure made screenings feasible and enabled medical providers to screen infants for phenylketonuria (PKU), a rare neurodegenerative metabolic disorder. In 1965 the American Academy of Pediatrics (AAP) Committee on the Fetus and Newborn recommended that every newborn in the United States be screened for PKU (Committee on Fetus and Newborn, 1965). Despite the recommendation of the AAP and the support of many advocacy groups for parents of children with mental handicaps, the medical community was slow to warm to the idea of universal screenings, in part due to the extremely low prevalence of PKU which is 1:15,000 (Therrell, 2001).

Research efforts in the 1960s focused on discovering screening methods for additional disorders; by the 1970s lab tests for SCD were developed, which incidentally identify carriers of the disease. In 1972, federal legislation known as the National Sickle Cell Disease Control Act was enacted, which provided the foundation for universal newborn screening for SCD. While speed of implementation varied across individual states, the National Sickle Cell Disease Control Act promoted universal screening as a way to decrease mortality and morbidity associated with SCD. In addition, it established counseling programs on a state-by-state level to serve the dual purpose of promoting public awareness of SCD and providing genetic counseling to individuals and families affected by the disease.

By 1974, several states had also passed legislation which mandated newborn screening to identify a variety of conditions, including SCD (Grover, Shahidi, Fisher, Goldberg, & Wethers, 1983). Despite advances in the science of newborn screening, many other state public health departments were slow to expand their newborn screening systems, in part due to the lack of a national newborn screening policy or models for guidance (Therrell, 2001).

## **Current State of Sickle Cell Screening**

Currently all states and the District of Columbia have universal newborn screening programs for SCD (National Newborn Screening and Genetics Resource Center, 2009). However, considerable variation exists among these programs in terms of which tests are utilized, which diseases are screened, and who is notified when a positive screening occurs (Stoddard & Farrell, 1997; Therrell et al., 2008).

A recent survey of sickle cell follow-up coordinators responsible for each state's universal newborn screening sickle cell programs was published in 2008 (Kavanagh, Wang, Therrell, Sprinz, & Bauchner, 2008). The authors were particularly interested in learning which stakeholders (i.e., primary care physician, hematologist, parent, etc.) were notified upon occurrence of a positive screening for SCD or SCT. All state sickle cell screening programs were found to notify the child's primary care physician (PCP) when the screening is positive for SCD (Kavanagh, et al., 2008). However, significant variation existed among which other stakeholders, if any, were also notified. Seventy-five percent of programs report the positive screening for SCD to hematologists and hospitals, while only 40% of state sickle cell screening programs notify families directly when a baby tests positive for SCD.

Significant variation exists across states for the number of agencies or individuals notified when SCT is detected in a newborn (Kavanaugh et al., 2008). Significantly fewer stakeholders are notified of a positive SCT result compared to a positive SCD result. Only 88% of screening programs notified the PCP when trait is identified; 63% notify hospitals, and only 37% directly notify parents by letter or phone call regarding the positive trait status of the child. Even more concerning is the finding that more than one-third of screening follow-up programs have no protocol in place when positive screenings for SCT occur.

## **Sickle Cell Screening in North Carolina**

In response to the passage of the National Sickle Cell Disease Control Act, the North Carolina Sickle Cell Program was established in 1973 to provide comprehensive services for individuals and affected families, as well as education and genetic counseling for the general public. Six comprehensive sickle cell medical centers exist across the state, which provide several services including medical treatment to adults and children with SCD, training for physicians and other medical providers on SCD, consultations with healthcare providers, and care via community clinics. Regional centers also frequently participate in national research studies investigating treatment options for SCD. The six comprehensive SCD centers in NC are located at Charlotte Medical Center, Duke University, East Carolina School of Medicine, Mission Hospital in Asheville, University of North Carolina at Chapel Hill, and Wake Forest University (North Carolina Sickle Cell Syndrome Program, 2009).

Infant screening for SCD began in NC in 1987 (Kirkman, 2008). However, at that time screening for hemoglobinopathies was limited to African-American infants, even though these conditions may be found in individuals from other ethnic backgrounds. North Carolina made sickle cell screening universal in 1994. State law GS 130A-125 now requires that a filter blood sample be taken from every newborn and submitted to the NC State Laboratory of Public Health (North Carolina Division of Public Health State Laboratory, 2009). If the initial screen is completed before 24 hours of age, a second test is required within 48-72 hours. The state laboratory in NC currently screens for eight “core” genetic conditions: congenital hypothyroidism, congenital adrenal hyperplasia, three types of SCD (HBSS, HBSC, and HBS-Beta thalassemia), biotinidase, transferase deficient galactosemia, and cystic fibrosis. In addition, newborns in NC are screened for 33 additional rare metabolic disorders (e.g., PKU,

Trifunctional protein deficiency, Homocystinuria, etc.; National Newborn Screening and Genetics Resource Center, 2009). When the NC laboratory identifies a positive screening for SCD, the laboratory notifies the child's PCP, the NC Sickle Cell Program, and a sickle cell counselor (Kavanaugh et al., 2008). When a positive screening for trait occurs, these same stakeholders are also notified. The director of the NC Newborn Screening Program also indicated that in NC a letter is sent to the mother of the affected newborn, informing her of the positive screening (S. Chaing, personal communication, May 17, 2010).

### **Disclosure of Sickle Cell Status**

As Kavanaugh et al. (2008) reported, state sickle cell screening programs vary significantly in who is notified of a positive screening for SCD or SCT, with primary care physicians (PCPs) being the most frequently notified stakeholder. When families are not notified directly by screening agencies, responsibility for communicating positive screening results typically falls upon PCPs, which can create a variety of problems. PCPs may have limited time for presenting educational information to families. PCPs also may not be able to immediately contact affected families. Listernick, Frisone, and Silverman (1992) reported that when sickle cell screening programs contact PCPs, there is frequently a significant delay in the families obtaining confirmatory diagnostic procedures. For this reason, the authors recommend forming a direct link between screening programs and families, rather than relying on an intermediary to convey such important information.

Another difficulty with relying upon PCPs to convey trait status is that they may be unprepared or unwilling to discuss genetic screening results with affected families. Such a conversation necessitates that the PCP provide accurate educational information as well as reassurance and comfort to families who may be in significant distress regarding the diagnosis.

In fact, when sickle cell follow-up coordinators were surveyed regarding the provision of counseling services following positive screening results, the sickle cell counselors reported that PCPs provided lower quality counseling than other professionals including subspecialty physicians, nurse specialists, and genetic counselors (Farrell, Certain, & Farrell, 2001).

### **Knowledge and Perception of SCD and SCT**

Few studies have examined public perception and knowledge of SCD, particularly among at-risk groups. In one of the few examples of systematic research into individual knowledge of sickle cell, Treadwell, McClough, and Vichinsky (2006) conducted focus groups and surveyed community members regarding perception and knowledge of SCT and SCD. This study was undertaken in response to a finding that in 2001 less than 18% of families in northern California who received notification that their infants were carriers of SCT took part in free SCT counseling. Therefore, researchers used focus groups to identify the barriers that may hinder families from pursuing counseling and other free services following a SCT disclosure.

Three focus groups were designed to represent various stakeholders: 1) healthcare providers, 2) individuals directly affected by the disease (e.g., individuals with SCD, family members of SCD patients), and 3) community members from predominantly African-American neighborhoods. It was found that members of all focus groups reported concern regarding the limited visibility and knowledge of SCD among the general public, emphasized the importance of using public awareness campaigns to educate community members, and highlighted that healthcare providers bear responsibility for educating patients about SCD and SCT. Some themes were not shared by all groups. The groups composed of individuals affected by SCD and African-American community members both reported that significant stigma is associated with the disease and that individuals with SCD and SCT frequently lack support from the medical

community. On the other hand, healthcare workers stressed the need for more outreach and education in the community, from grade school to college. This discrepancy among focus groups serves as a cautionary tale to those in the health field. While healthcare workers may view SCD from a medical model, those who are most affected by the disease may view SCD or SCT as taboo conditions and be resistant to counseling and education initiatives.

Ogamdi (1994) investigated knowledge of SCD by surveying 334 college students at a predominantly African-American university in southeastern Texas. The sample was unique in that it consisted predominantly of black individuals in their prime reproductive age (early 20s); the sample was chosen because the researcher believed this group to be most in need of education and knowledge regarding responsible reproductive choices given the high prevalence of SCD and SCT in African-American communities. The survey consisted of eight questions that surveyed for demographic information, five questions designed to obtain epidemiologic information, and 15 multiple choice questions which assessed knowledge of critical aspects of SCD.

Results indicated that the basic facts of SCD were not well understood by individuals most likely to be directly affected by SCD and SCT (African-Americans). Only 55% could correctly identify which cell in the body becomes sickled and nearly a third (28%) were unaware of how the disease is transmitted (i.e., genetic inheritance). Regarding symptoms and complications of SCD, 37% of the African-American university students did not know the most frequent symptoms, such as fever, pain crises, swelling, and anemia. More than 60% did not know that SCD could be prevented through responsible reproductive practices. Only 40% understood that because of added risk, those who carry SCT should consult a doctor regarding

pregnancy planning. Finally, many of the students in the study were unable to recall whether they had been tested for SCT or SCD.

Knowledge of sickle cell and disclosure patterns were further examined by Acharya, Walsh Lang, and Friedman Ross (2009) who surveyed a targeted population consisting of parents of children with SCD, parents of children with SCT identified through newborn screening, and parents with SCT who were uncertain of their children's status. Researchers recruited ( $n = 53$ ) participants through several Chicago sites including a sickle cell disease clinic, the Sickle Cell Disease Association of Illinois, a public health clinic, and a postpartum inpatient unit. The majority of participants reported having SCT and approximately half ( $n= 27$ ) had a child with SCD. Participants underwent structured interviews to gather the SCT status of respondents and their children, disclosure patterns and intent, knowledge of SCD, sources of knowledge, health orientation to SCT, and perceived stigma and experiences of discrimination as related to SCT status.

Knowledge of SCD was measured by 10 true-false questions about sickle cell inheritance and the meaning of screening results. Most knowledge items were adapted from a survey administered to Nigerian college graduates (Adewuyi, 2000; as cited in Acharya, Walsh Lang, & Friedman Ross, 2009, p. 1164). Questions assessed inheritance patterns of SCD (i.e., Sickle cell disease can be inherited if one parent has the trait and the other parent does not. [False]), as well as understanding of sickle cell screening results (i.e., A negative sickle cell carrier test means: a) s/he is very unlikely to have the trait or b) s/he definitely does not have the trait. [b]). Sources of knowledge of sickle cell were assessed by providing participants with a list of 15 sources; respondents were instructed to select all that applied. Participants were asked about their intent to disclose their own SCT status to their children. If they had children with SCT, participants

were also asked about whether and when they planned to disclose their children's SCT status to them.

Results indicated that there was significant misunderstanding about how SCD is inherited, with a mean score of 68% on the 10-item true/false knowledge questionnaire. Participants who had a child with SCD scored significantly higher than those without a child with SCD on the knowledge questionnaire (78% vs 58%,  $p = .002$ ). The most common sources of knowledge for parents of a child with SCD were pediatricians (89%), SCD clinic staff (89%), and employees of the Sickle Cell Disease Association of Illinois (78%). For those without a child with SCD, the most common sources of knowledge included family members (63%), ob/gyn providers (42%), and pediatricians (38%). Parents who had a child with SCD were significantly more likely to have received information from formal educational sources or health care-related sources and to have researched SCD using the Internet than those without a child with SCD. Very few participants reported having received information from genetic counselors.

Most parents indicated a desire to disclose both their SCT status and to inform children of their SCD and SCT statuses. Of the 47 parents with SCT, only one indicated that she had not disclosed her SCT status to her children and would not disclose her status. Four did not reply to the question about personal SCT disclosure. Of the 58 children with SCD and SCT, 45 children, consisting of 18 with SCT and 27 with SCD, had been told of their status or would be told in the future. Most children (84%) learned or would learn about their sickle cell status prior to the age of 13.

### **Stigma Associated with Sickle Cell Trait**

In addition to assessing sickle cell knowledge and disclosure intent among parents with SCT, Acharya, Walsh Lang, and Friedman Ross (2009) also assessed perceived stigma

associated with sickle cell carrier status. Forty-seven adults with SCT completed a standardized stigma scale designed to measure stigma related to HIV, which consisted of 8 items on a 5-point Likert scale. Participants also responded to three questions about insurance and employment discrimination. Results from 43 participants were included in the analysis; the average stigma score per item was less than 2 ( $M = 1.6$ ; scale from 1-5 with 5 indicating high stigma), suggesting a low sense of stigma associated with carrier status. The stigma statement most frequently “agreed” with, “I am very careful who I tell that I have sickle cell trait,” was endorsed by 10 of the 43 respondents (24%). All participants disagreed or strongly disagreed with the statement “I worry about people discriminating against me.” Two of the 47 carriers reported that they had either been denied insurance or had been offered insurance at a prohibitively high cost due to their positive trait status. No respondents reported experiencing employment discrimination based on their SCT status.

In contrast, semi-structured interviews conducted with women with SCT suggest that positive carrier status may be stigmatizing for some individuals. Asgharian and Anie (2003) conducted interviews with 35 British women of African or African-Caribbean origin. Women reported significant anxiety disclosing their trait status to partners in advance of becoming pregnant. Women with SCT indicated that it was often easier to risk giving birth to an affected child rather than to initiate potentially uncomfortable conversations with partners and risk rejection. By coding interview transcripts, researchers identified several obstacles which frequently hindered women with SCT from discussing their status with partners. Women were reluctant to initiate a dialogue which was frequently perceived to be awkward and sensitive. Their own lack of knowledge regarding SCD and SCT status often contributed to this apprehension. Timing of the discussion within the context of a relationship was also noted to be

difficult. Women with trait indicated that revealing their status “too soon” in the relationship could cause problems, as highlighted by one woman’s response, “Once you have chosen your partner and are dating, at what point in the relationship do you ask them to do a blood test?” (Asgharian & Anie, 2003, p. 29). Conversely, some women also expressed concern that waiting too long in a relationship before disclosing SCT status could cause their partners to feel deceived.

Fifteen of the 35 women interviewed did not find out their partner’s status prior to conception. Some of these women reported that they in fact had actually “forgotten” about their own status, until they reached a point in their lives in which they were either already pregnant or considering becoming pregnant. In such cases, Hill (1994) suggests that the women may in fact be engaging in “obfuscation,” in which they subconsciously confuse or even deny information regarding their own medical status that may threaten their own reproductive autonomy.

Many women indicated that they perceived significant stigma was associated with being a trait carrier, and thus were hesitant to reveal information to partners and face rejection. Researchers also cited ignorance as a key factor in the lack of open communication regarding women’s trait status. Not all participants had a clear understanding of what having SCT actually meant nor what risks to offspring were. In addition to a general lack of sickle cell knowledge, the age at which participants had learned their own trait status was highly variable. Unfortunately, the authors did not report the ages at which women became aware of their status.

### **Sickle Cell Trait Counseling**

One mechanism which may be particularly helpful both to combat misinformation regarding SCT and to reduce stigma is the provision of counseling post SCT disclosure. Individuals with SCT or parents of an infant who has tested positive for SCT should be provided

with an opportunity to speak to a trained counselor who is familiar with SCD and SCT.

Counseling post SCT disclosure should fulfill several primary purposes: 1) to educate regarding biological aspects of SCD and SCT including the mechanism of genetic transmission, 2) to correct any misconceptions, and 3) to alleviate potential psychosocial complications that arise with disclosure (Headings & Fieldings, 1975). Common misconceptions include that SCT frequently causes medical problems and results in many deaths each year. In addition, some affected individuals may fail to understand the genetic nature of sickle cell and instead believe that SCT is transmitted through sexual contact or is a contagious disease similar to measles (Headings & Fielding, 1975).

SCT disclosure occurs primarily to ensure that affected individuals are aware of the potential risk of genetic transmission to any future children. Therefore, sickle cell counselors must be able to clarify the role of genes in the transmission of SCT, discuss the risks and probability of having children with trait or disease, and explain options for preventing the occurrence of SCD. Headings and Fieldings (1975) outline several possible options which could be presented to adolescents if they desire to prevent the transmission of the sickle cell gene to any future children, including: 1) engage in sexual activity without regard for risk, 2) select a partner who does not have SCD or SCT, 3) adopt children in the future, and 4) choose to remain childless. With the advances in reproductive technology which have occurred in the past decade, couples now may have additional options including pre-implantation genetic diagnosis (PGD), in which testing is completed to determine whether an embryo carries a particular gene prior to implantation via in vitro fertilization.

## **Need for Education**

Results of surveys on sickle cell knowledge (e.g., Ogamdi, 1994), indicate that basic facts regarding sickle cell and its transmission are not well understood even by those who are most likely to be affected by it. As Ogamdi (1994) summarizes:

Clearly, universities and colleges, especially those with a student body that is predominantly Black, should make sickle cell screening, education, and counseling available to all at the time of enrollment. Those who discover that they carry the trait will then be aware of the factors they must consider in making reproductive choices and will be better able to deal with problems that may arise should they become parents of a child with sickle cell disease. (p. 236)

While Ogamdi's (1994) study evaluated knowledge of SCD and SCT among college-aged students, high rates of sexual activity among teenagers necessitate that sickle cell awareness campaigns extend to high schools and particularly emphasize increasing knowledge of SCD and SCT among African-American adolescents. Every two years, the Centers for Disease Control and Prevention collect data on adolescent health behaviors through the Youth Risk Behavior Surveillance System (YRBSS). The YRBSS is a national school-based survey which asks adolescents to self-report on a variety of health behaviors. Data from the 2007 YRBSS indicates that over 52% of high school students in N.C. have had sexual intercourse (CDC, 2009). African-American students report much higher rates of sexual intercourse, with nearly 66.5% of the national sample of African-Americans reporting having had sex, compared with only 43.7% of white students. In addition, 16.3% of African-Americans reporting engaging in sexual intercourse prior to age 13, compared with only 4.4% of white students (CDC, 2009). N.C. has consistently battled high teenage pregnancy rates and ranked ninth in the country for teen pregnancy in 2007 (NC Healthy Schools, 2009). African-American adolescents have particularly high rates of pregnancy. Data from 2007 indicates the pregnancy rate for African-American girls between the ages of 15 and 19 was 87.1 out of 1000 (NC Healthy Schools, 2009).

The YRBSS does provide some information on safe sex behaviors, though this information is not broken down by ethnic background. In 2007, among all sexually active youth, only 61.5% of teenagers reported using a condom the last time they had sex and just 16% of sexually active adolescents reported being on birth control pills (CDC, 2009). Therefore, African-American adolescents are at high risk of engaging in unprotected sex and may be unaware of their own SCT status, thus increasing the likelihood of conceiving children that carry SCT or have SCD.

### **Purpose of the Current Study**

Research is lacking about level of knowledge of SCD and SCT among the population most likely to be directly affected by this condition in the U.S. (i.e., young African-Americans). Only a few studies have examined how familiar African-American adults are with SCD, and these studies suggest that there is significant misinformation among African Americans regarding the transmission, symptoms, and treatment of SCD. Only one published study has assessed level of knowledge of SCD among African-American college students (Ogamdi, 1994). This study indicated low levels of knowledge about SCD. Only descriptive statistics were reported, and the study did not assess knowledge of sickle cell trait. No studies have used measures specifically designed to assess knowledge of SCT even though far more individuals are affected by SCT than SCD. Furthermore, no studies have assessed whether knowledge of disease or trait status is predictive of increased sickle cell knowledge.

While controversy has existed over whether positive screenings for SCT should be disclosed to the families of affected newborns given the possibility of negative outcomes (e.g., anxiety, disclosure of non-paternity), currently the vast majority of states do alert some stakeholders (e.g., primary care physicians, parents) when a newborn tests positive for sickle cell trait (Kavanagh et al., 2008). Such a disclosure is made in an effort to empower individuals and

families by equipping them with knowledge which may help them in making future health and reproductive choices. A line of research has examined how medical providers and state sickle cell programs transmit information regarding a child's SCT status to parents of the affected child and potential risks and benefits of such transmission. However, no studies have yet examined the secondary route of transmission (i.e., from parent to child) to identify whether information is in fact being passed down to the affected individual.

The information that is transmitted from parent to child regarding a child's health condition is particularly important in SCT, as the presence of SCT typically has few implications for an individual's health, but becomes critically important as an individual reaches reproductive maturity. Because a person with SCT has a 25% chance of having a child with SCD if the other parent also has trait and a 50% chance of passing on the trait to their children regardless, information regarding genetic inheritance, risk of SCD, contraception, and reproductive options should be shared with individuals with SCT prior to them becoming sexually active. How adults initiate such a conversation, what information is communicated, and when this information is shared have important implications not only for the individual, but also for children which may be conceived in the future. Such information is particularly critical during adolescence and young adulthood, since this is the developmental period in which a majority of individuals become sexually active. This time period is also critical due to the rare but serious potential complication of sickle cell trait sudden death, which may result from participation in high school and college sports.

Because sickle cell screening began in NC in 1987 for all African-American infants, the current generation of young adults is the first wave of individuals for whom SCT status was established and disclosed at birth. African-American young adults in NC who have SCT should

ideally be aware of their positive trait status since their families were notified at birth following the positive screening. However, no studies have evaluated whether these individuals are aware of trait status.

The purpose of the present study was first to examine knowledge of SCD and SCT among African-American college students. It is hypothesized that they will possess low levels of general knowledge about SCD and SCT. It is also hypothesized that there will be low levels of knowledge regarding the genetic transmission of SCD and SCT and the potential health implications of the conditions.

A second purpose was to examine participants' knowledge of their own SCT status. It is hypothesized that a significant number of participants will be uncertain about their trait status. While 8% of African-Americans carry SCT, many of these individuals may be unaware of their positive trait status. Though nearly all U.S. states identify individuals with SCT at birth, no studies have yet examined whether this information is accurately transmitted to the individual, whether by a family member or healthcare provider, prior to adulthood. If individuals with SCT have not been informed of their trait status, they will likely assume that they do not have trait since the condition is typically asymptomatic.

The study also examined whether a link exists between SCT status and sickle cell knowledge. It is hypothesized that awareness of SCT status will be positively correlated with knowledge of SCT; thus individuals who know that they possess SCT will have more knowledge of SCT than individuals who do not have SCT or are unaware of their trait status. It is also hypothesized that individuals with a family member who has SCD will have more accurate sickle cell knowledge than young adults who do not have a family member with SCD.

The final exploratory purpose is to examine the sources of SCD and SCT knowledge among African-American college students. It is hypothesized that among individuals who have some knowledge of sickle cell, those who have received information about SCT and SCD from family members will demonstrate increased knowledge compared to those who have received information from media, friends, school, or health care professionals.

### **Hypotheses**

Thus, four a priori hypotheses were developed to examine knowledge of sickle cell trait and disease among African-American college students:

1. Participants will possess low levels of overall knowledge of SCT and SCD, including misinformation regarding genetic transmission and common symptoms.
- 2a. A significant number of participants will be uncertain of their sickle cell trait status.
- 2b. There will be a significant number of false negatives or individuals who believe they do not have SCT when in fact they do.
3. Knowledge of positive SCT status will be positively associated with increased sickle cell trait knowledge.
4. Participants who have received information on sickle cell from family members will demonstrate increased knowledge over those who have received information from media, friends, teachers, or health care professionals.

## Chapter II: Method

### Participants

A total of 269 undergraduate students from East Carolina University originally enrolled in the study. At the time of the study, participants were taking an introductory psychology course, which required completion of a research participation assignment. The assignment may be fulfilled in several ways, including spending a total of five hours participating in a research study in the department of psychology. Participants received credit for 30 minutes of research participation in the current study. Participants were recruited through Experimentrak, an online system for collecting demographic data for the purpose of identifying potential subjects and for collecting research data. One of the demographic questions that student participants must answer when creating an account through the Experimentrak system is “What is your race/ethnicity?” Seven responses are provided: 1) American Indian/Alaska Native, 2) Asian, 3) Black/African American, 4) Hispanic/Latino, 5) Native Hawaiian/Pacific Islander, 6) White, and 7) Other. Only students who selected “Black/African-American” as their ethnicity were eligible to participate in the current study.

Two of the 269 participants exited the study mid-session. They were provided with debriefing information by the researcher via email, and their data were excluded from the study. One additional participant was excluded because the participant provided contradictory information regarding his or her ethnicity. While the participant’s Experimentrak account indicated that his or her ethnic background was “Black/African American,” the participant selected “Caucasian” when answering demographic questions for the current study. Six participants were excluded because they were over 30 years old and the purpose of the study was to explore knowledge of sickle cell among college-aged individuals in their teens and twenties.

Finally, two participants were excluded for providing contradictory information regarding their SCT and SCD status. Both participants reported being positive for both SCT and SCD and thus were excluded from the study.

This resulted in a final sample size of 258 participants. Participants ( $N = 258$ ) ranged in age from 17 years, 10 months to 29 years, 10 months, with a mean participant age of 19 years, 4 months ( $M = 232.2$  months;  $SD = 18.3$  months). There were more female participants (67%,  $n = 174$ ) than male participants (33%,  $n = 84$ ). Most participants indicated that their ethnic background was “African-American” (96%), while 10 reported their ethnicity to be “Biracial” (4%), and one reported “Other” (<1%). A majority of participants (66%,  $n = 169$ ) reported that they were born in North Carolina, while 34% ( $n = 89$ ) indicated that they were born elsewhere. Of those who were born elsewhere, most (31%,  $n = 80$ ) reported having been born in a different state, while nine (3%) indicated that they were born in another country. Two participants each reported that they were born in Germany, Sierra Leone, and Liberia. One participant each reported having been born in Haiti, Canada, and Nigeria. Five participants (2%) indicated that they were parents, a fact which may confer more knowledge about genetic testing for newborns.

## **Measures**

**Demographic Variables.** Participants provided demographic data (Appendix A) including gender, ethnic background, and whether they had any children. They also reported other variables such as place of birth that would have been needed to access individual newborn screening results in the NC State Laboratory of Public Health’s online database.

**Personal and Family Health Questionnaire.** A personal and family health questionnaire was created for this study (Appendix B). This multiple choice survey asks whether the participant has previously heard of the terms “sickle cell disease” and “sickle cell trait.” The

measure contains two questions to assess participant's disease and trait status. It provides five options: "Yes," "I think I do," "I don't know," "I don't think I do," and "No." Two questions then assess whether anyone in the participant's family has SCT or SCD. If the participant indicated having a family member with SCT or SCD, he or she was provided with several options to identify affected family members. Options were: mother, father, sister, brother, daughter, son, aunt/uncle, cousin, grandmother/grandfather, and other. The measure also asks the participant to identify where he or she has learned about sickle cell. Five knowledge sources were provided: 1) At school, 2) From family, 3) From friends, 4) From a doctor or nurse, and 5) From the TV, movies, or something you read. The participant could also select "Other" as a source of knowledge. Those who selected "other" were asked to provide the other source of knowledge.

**Sickle Cell Trait Knowledge Questionnaire.** A measure was created for the current study to assess participant knowledge of SCT (Appendix C). The Sickle Cell Trait Knowledge Questionnaire (SCTKQ) is a 12-item true/false test which asks about general SCT knowledge, including genetic transmission, general health of individuals with SCT, and newborn screening. Items were generated by a team of sickle cell researchers, including the author. Several questions were designed to assess whether misconceptions exist regarding SCT (e.g., SCT frequently causes health problems). Reliability of the SCTKQ was evaluated as part of the current study.

**Transition Knowledge Questionnaire.** The Transition Knowledge Questionnaire (TKQ; Appendix D) is a 25-item tool that was designed to assess knowledge of SCD in seven areas: 1) pathophysiology, 2) genetics, 3) physical manifestations, 4) treatment, 5) self-care, 6) psychosocial and developmental issues, and 7) health care delivery system (Newland, Cecil, &

Fithian, 2000). The TKQ assesses sickle cell knowledge using multiple choice and true/false questions. Higher scores on the TKQ represent greater knowledge, with a score of 25 being the total possible points. The measure was developed to assess knowledge of SCD among adolescents with SCD prior to their transition from pediatric clinic care to adult care. Researchers originally generated a 50-item question pool. Through content validity analysis, expert evaluation for relevance and clarity of questions, and reliability evaluation, the pool was reduced to 25 final questions with one point awarded for each correct answer. Reliability of the TKQ was assessed by administering the scale to nine adolescents with SCD who ranged in age from 8<sup>th</sup> grade through high school graduate. Chronbach's alpha was .79, establishing sufficient internal consistency though the sample size was quite small. Readability analysis indicated that the TKQ is on a 6<sup>th</sup> to 7<sup>th</sup> grade reading level (Newland, Cecil, & Fithian, 2000).

Newland (2008) later used the TKQ to assess knowledge of SCD among a sample of 74 adolescents with SCD between the ages of 14 and 21 in order to identify factors that predict successful transition from pediatric to adult care. For this sample, total score on the TKQ ranged from 9 to 25 ( $M = 18.58$ ,  $SD = 3.55$ ; Chronbach's alpha = .71; Newland, 2008). There were no statistically significant differences between males and females, and no questions were answered correctly by all participants. Participants scored higher on questions assessing psychosocial/developmental issues and healthcare provider issues and lower on questions involving genetics, inheritance, and treatment options (Newland, Cecil, & Fithian, 2000).

The TKQ was modified for the current study, resulting in the Revised Transition Knowledge Questionnaire (RTKQ; Appendix E). A prompt was added which tells participants that the questionnaire is about SCD. Questions 7, 10, 15, 19, 20, 21, 24, and 25 were eliminated because they required an advanced knowledge of treatment that would be relevant to individuals

with SCD but not appropriate for a general knowledge measure (e.g., Before going to the Emergency Room, a patient [with SCD] should notify his/her regular doctor. True or False). Question 8 was altered to ask about disease rather than trait so that the measure would focus exclusively on SCD. A question was also added (question 19 on the RTKQ) to assess whether participants are familiar with the most common symptom of SCD (i.e., pain crises). Reliability of the RTKQ was evaluated as part of the current study.

### **Procedure**

Institutional Review Board approval was obtained for the study (Appendix F). Undergraduate students enrolled in introductory psychology courses were recruited through Experimentrak, an online research participation system. Only students who identified themselves as “Black/African American” when creating an Experimentrak account were eligible to participate in the study. Recruitment of participants took place between April 2010 and December 2010. All data were collected through Experimentrak. After selecting the study from a list of available research opportunities, participants were provided with informed consent materials (Appendix G). The consent process outlined the purpose and length of the study, what participation entailed, possible risks and benefits, and limits of confidentiality. Participants were informed that they would be asked to provide some background and personal health information and then to complete four online questionnaires. Participants were informed that since 1987, North Carolina has tested newborns for the presence of sickle cell. Participants were informed that by agreeing to participate in this study, they gave permission for the researchers to use the demographic information they provided to access results of newborn screenings in the NC State Laboratory of Public Health database. Two participants, who after reading the informed consent,

decided not to participate, were able to exit the online system at that time prior to providing any personally identifiable information.

Participants who provided consent were directed to the first of the four study questionnaires and proceeded through each measure. Participants who wished to discontinue participation were able to exit the study at any time by closing their browser. After completing the four online questionnaires, participants were shown a screen informing them that they had the option to receive a summary of research findings as well as any information that was accessed about them from the NC State Laboratory of Public Health. Participants answered “yes” or “no” to indicate whether they wanted a summary of findings and whether they wished to be contacted regarding their trait or disease status. These two questions failed to be posted online prior to completion of the study by the first 12 participants. These participants were contacted individually by the researcher via email and provided with the opportunity to request a summary and any personal health information found in the database.

The final screen contained debriefing information (Appendix H) which explained the purpose of the study a second time and provided contact information (email and telephone number) for the principal investigator if participation in the study caused discomfort or distress. Additional contact information was also provided for the university counseling center, the Sickle Cell Disease Association of American (SCDAA), and the university institutional review board as resources in case of distress, for more information about sickle cell, or to report any concerns about the study, respectively.

Appropriate measures were taken to protect confidentiality. All data uploaded from the Experimentrak site were kept in a password protected Excel file. Any printed data were kept in a secure, locked filing cabinet. Only the researcher and the primary investigator had access to the

contents of the data files. After the participation window was closed, researchers accessed the NC State Laboratory of Public Health database to locate participants' newborn screening results in order to verify SCT and SCD status. Researchers searched the database for all participants who reported having been born in NC, using demographic variables including date of birth, place of birth, name at birth, and mother's maiden name. However, no newborn screening results existed in the database for any participants. The director of the state laboratory's newborn screening program, Dr. Sue Chaing, was contacted and indicated that in the early years of newborn screening, the state laboratory's policy was to discard lab results after two years. Dr. Chaing indicated that some lab results from the 1990s may have been preserved on microfiche, but only in 1996 did the state laboratory begin to store records electronically (S. Chaing, personal communication, May 17, 2010). Directors of the NC Sickle Cell Disease Association were also contacted as to where such records may be (M. Wright & D. Marris, personal communication, May 27, 2011). They suggested that newborn screening results prior to the mid-1990s may have been sent to individual public health departments.

In addition to missing records, the public health database also was found to be cumbersome to search. Four pieces of information were required to search for any individual's records. If errors or omissions were made in any field, records could not be accessed. It was discovered that actual newborn screenings results are not freely accessible through the database. Instead, a researcher must request that screening results be "released" from the entity which originally requested them.

Prior to data analysis, participants' names were removed from the data set and replaced by a three-digit participant ID number.

## **Data Inspection and Analyses**

**Data Screening.** The first step in data analysis involved screening the data for violations of normality and for missing values. Data screening was conducted using Microsoft Excel and SAS 9.2. During this step, one participant was identified as having reported his or her race/ethnicity as Caucasian and was excluded from the study. Six participants were identified who were over 30 years of age and were excluded from the study. Two participants were identified who answered “yes” to both “Do you have sickle cell trait?” and “Do you have sickle cell disease?”. They were also excluded from the study. A total sample size of 258 participants remained, which consisted of 174 females and 84 males.

**Missing Data.** Data were then screened for missing values. Ten of the 12 participants whom the researcher contacted to provide the final two questions of the study did not reply to indicate whether they wished to be provided with a summary of study results or to be notified if their trait status information was found in the state laboratory database. For the variable of “birthdate,” six responses were determined to be likely invalid because the year provided for “year of birth” was either too old (i.e., 1900) or too young (i.e., 1997, 2010, 2011). These questionable responses were treated as missing data. One questionable response for “birthplace” was identified. One participant responded “town.” This response was deleted and treated as missing data.

## **Statistical Analyses**

SAS 9.2 was used to conduct all statistical analyses. The criterion for significance tests for all *a priori* hypotheses was set at  $\alpha = .05$ .

Prior to data analyses, Chronbach’s alpha was obtained for the Sickle Cell Trait Knowledge Questionnaire (SCTKQ) and the Revised Transition Knowledge Questionnaire

(RTKQ) to verify sufficient levels of reliability. An iterative process was used to eliminate items with low item-to-total correlations to improve reliability. For the SCKTKQ, questions 5 and 11 were selected for elimination. Question 5 (People with [SCT] who are athletes such as football players may be at-risk for sudden death: True/False) had a negative correlation with total ( $R = -.37$ ), and its exclusion raised alpha from .43 to .56. Question 11 (All states in the U.S. screen newborn babies for [SCT] as well as other conditions: True/False) had a very small correlation with total ( $R = .02$ ). Its exclusion raised alpha from .56 to .59. For the RTKQ, questions 4 and 6 were selected for elimination. Question 4 (People with [SCD] have mainly hemoglobin \_\_\_ in their red blood cells: A, B, S, or E) had a small correlation with the total ( $R = .11$ ) and its removal raised the measure's alpha from .62 to .63. Question 6 (Hemoglobin SC disease and sickle beta thalassemia are forms of [SCD]: True/False) also had a small correlation with the total ( $R = .03$ ). Its exclusion raised alpha from .63 to .65.

Factor analyses were conducted on both the Revised Transition Knowledge Questionnaire and the SCT Knowledge Questionnaire. However, factor analysis failed to identify either a distinct factor or multiple factors underlying the scales.

## Chapter III: Results

### General Knowledge of SCT and SCD and Self-Reported Status

“Sickle cell trait” is a term that was familiar to most of the sample with 91% ( $n = 236$ ) reporting that they had heard of the term before, as shown in Table 1. As anticipated, a large percentage of participants were not certain of their own personal trait status. There were 5% who indicated that they have SCT ( $n = 14$ ), and 42% who indicated that they do not have SCT ( $n = 108$ ). However, 14% ( $n = 36$ ) responded “I don’t know,” 36% ( $n = 94$ ) responded “I don’t think I do,” and 2% ( $n = 6$ ) responded “I think I do.” Together, the three groups who indicated uncertainty about their personal trait status comprised 52% of the sample.

When asked whether any family members have SCT, 16% ( $n = 40$ ) stated “yes,” and 22% ( $n = 57$ ) stated “no.” The majority of participants (62%,  $n = 161$ ) reported “I don’t know.” When asked how many family members have SCT, 11% of participants ( $n = 29$ ) reported having one family member with SCT. Seven participants (3%) reported having two family members with SCT, four had three family members (2%), and one participant (<1%) reported having five family members with SCT.

The overwhelming majority (97%,  $n = 250$ ) of participants indicated that they had heard of the term “sickle cell disease,” as shown in Table 1. In terms of disease status, a small but substantial portion of the sample was not certain about whether they themselves had SCD. Two percent of the sample ( $n = 5$ ) reported that they have SCD, and 77% indicated that they did not have SCD ( $n = 199$ ). However, almost one in five (18%,  $n = 46$ ) selected the response “I don’t think I do,” and another 3% ( $n = 8$ ) responded “I don’t know” when asked if they had disease.

In terms of family members with SCD, 10% ( $n = 27$ ) of participants indicated having a family member with SCD, while 36% ( $n = 92$ ) responded “I don’t know,” and 54% ( $n = 139$ ) responded “no.” Nine percent of the sample ( $n = 23$ ) indicated that one family member has

SCD, while two participants (1%) reported that two family members have SCD, and one participant (<1%) reported that three family members have SCD.

### **Knowledge Sources**

Participants were asked to report where they had learned about sickle cell. Slightly more than 3% ( $n = 9$ ) indicated that they had not received any information about sickle cell previously. The average number of knowledge sources was two ( $M = 2.11$ ,  $SD = 1.25$ ). Most participants had learned about sickle cell from one source (34%,  $n = 87$ ) or two (33%,  $n = 84$ ) sources. Fifteen percent of the sample reported having learned about sickle cell from three source ( $n = 39$ ). Smaller proportions reported more sources of sickle cell knowledge: four (10%,  $n = 25$ ), five (5%,  $n = 12$ ), and six (1%,  $n = 2$ ).

The most frequently endorsed source of knowledge was school, with 82% ( $n = 212$ ) of participants indicating that they had received sickle cell knowledge from school. The second most frequently endorsed source of knowledge was the media with 42% ( $n = 108$ ) of participants indicating that they had learned about sickle cell from television, movies, or something they had read. Family was endorsed as a knowledge source by 30% ( $n = 78$ ) of the sample. Health care providers (doctor/nurse) as a knowledge source was endorsed by 21% ( $n = 54$ ), and 20% ( $n = 51$ ) of participants reported having learned about sickle cell from friends. Fourteen percent ( $n = 35$ ) indicated that they had received information from another source. Some “other” sources listed by participants included pamphlets, research, a health convention, athletic activity, football physicals, sickle cell informational society, and counselor.

### **Preference for Personal Trait Status Information & Research Summary**

When offered the opportunity to learn about their personal sickle cell trait status, 44% ( $n = 108$ ) indicated that they did not wish to be informed about their status if the information was

available in the NC State Laboratory of Public Health. A little less than half of the sample (48%,  $n = 120$ ) indicated that they wanted to be provided with a summary of research findings upon the study's completion.

### **Internal Consistency of Scales**

To assess the internal consistency of both knowledge scales, Cronbach's alpha was run. An iterative procedure was used to identify questions whose removal would improve reliability. After a question was removed, alpha was calculated for remaining questions. Using this method, two items from each scale were selected for removal because they were not highly correlated with the rest of the scale items. From the Sickle Cell Trait Knowledge Questionnaire (SCTKQ), questions #4 and #11 were deleted, resulting in a 10-item scale. Deleting these questions from the total scale improved alpha from 0.43 to 0.59. On the Revised Transition Knowledge Questionnaire (RTKQ), questions #4 and #6 were deleted, resulting in a 16-item scale. Deleting these questions from the total scale improved alpha from 0.62 to 0.65.

### **Hypothesis #1**

There was significant misunderstanding regarding SCT. Scores on the ten question, true/false SCTKQ measure ranged from 0 to 10 ( $M=6.80$ ,  $SD=2.05$ ), with an average of 68% correct. Questions which were incorrectly answered by over 25% of participants were individually reviewed. Of the 258 participants, 142 or 55% incorrectly believed that sickle cell trait can "turn into" sickle cell disease. Ninety-six or 37% believed that sickle cell trait causes lots of pain crises. Over a quarter of respondents (26%,  $n = 66$ ) did not believe that most people with sickle cell trait live long, healthy lives. Over a quarter of respondents (26%,  $n = 67$ ) believed that if a person has sickle cell trait, all of his or her children would also have sickle cell trait. Seventy-eight participants or 30% of the sample were unaware that people from all

racess/ethnic backgrounds can be affected by sickle cell trait. A majority of participants (58%,  $n = 150$ ) believed that sickle cell trait causes many medical problems for most individuals.

Finally, 107 participants (41%) believed that sickle cell trait results in many deaths per year.

Though question five was excluded from the final analysis of the SCTKQ, a majority of participants incorrectly answered it. Thus descriptive statistics will be reported. One-hundred and thirty five participants (52%) were unaware that “people with sickle cell who are athletes such as football players may be at-risk for sudden death.”

Participants performed better on the RTKQ, which evaluated knowledge of sickle cell disease. The final version of the RTKQ consisted of sixteen questions, including 13 four-option multiple choice questions and three true/false questions. Scores on the revised RTKQ measure ranged from 2 to 16 ( $M=12.15$ ,  $SD=2.45$ ), with an average of 76% correct. Questions which were incorrectly answered by over 25% of participants were individually reviewed. Of the 258 participants, 104 participants (40%) were unaware that people with SCD inherit two genes for sickle hemoglobin, one from the mother and one from the father. A total of 113 participants (44%) were unaware that children with sickle cell disease take penicillin daily to decrease risk of serious infection. A large portion of participants were unfamiliar with sickle cell symptoms which occur during adolescence: 26% ( $n = 67$ ) did not know that physical development in adolescence may be delayed, and 32% ( $n = 83$ ) did not correctly identify that adolescents with SCD may be shorter than others their age, tire more easily, and mature later than peers. A substantial portion of participants were unfamiliar with certain complications of sickle cell: 39% ( $n = 100$ ) did not identify priapism, acute chest syndrome, and leg ulcers as being common symptoms of SCD and 35% ( $n = 90$ ) were unable to identify the most common symptom of SCD

(i.e., pain episodes). In addition, nearly half (48%,  $n = 124$ ) were unaware that individuals with SCD may be able to exercise or play sports safely.

Though it was excluded from final analysis, descriptive statistics will be reported for question 4 on the RTKQ. A majority of participants (64%,  $n = 164$ ) were unable to identify the type of hemoglobin individuals with SCD possess (i.e., S). A significant correlation existed between total trait knowledge and disease knowledge ( $r = 0.35, p < .0001$ ).

### **Hypothesis #2a**

As hypothesized, a significant portion of the sample was uncertain of their SCT status. When asked “Do you have sickle cell trait?”, a majority of respondents (52%,  $n = 136$ ) indicated that they were not certain of their trait status by responding either “I don’t know” (14%,  $n = 36$ ), “I don’t think I do” (36%,  $n = 94$ ), or “I think I do” (2%,  $n = 6$ ). The second largest group responded “No” (42%,  $n = 108$ ), and a small portion of the sample indicated “Yes” (5%,  $n = 14$ ).

### **Hypothesis #2b**

Because newborn screening results could not be located, hypothesis 2b was unable to be tested, and it is unknown whether any participants reported that they do not have SCT when in fact they do or, conversely, reported that they do have SCT when they do not. However, national epidemiologic prevalence rates for SCT were reviewed and compared to reported trait percentage. Prevalence rates of SCT in the United States are estimated to be 8% among African-Americans (Tsaras, Owusu-Ansah, Owusua-Boateng, & Amoateng-Adjepong, 2009). In the current study, 5% of participants indicated that they had SCT.

### **Hypothesis #3**

Participants’ self-reported trait statuses were unable to be verified. Thus hypothesis #3 could not be tested as written (i.e., comparing total trait knowledge scores of those who know

they have SCT with those who do not know they have SCT). Thus the decision was made instead to compare total trait knowledge scores of participants who reported having SCT with total trait knowledge scores of participants who did not report having SCT. Participants who reported having SCT ( $n = 14$ ) differed significantly from participants who reported not having SCT ( $n = 244$ ) in their SCT knowledge. Total SCKQ score was significantly higher for participants with SCT ( $M = 8.57$ ) than for those without SCT ( $M = 6.70$ ),  $t(17.5) = -5.31, p < .0001$ .

Participants who reported having SCT ( $n = 14$ ) did not differ from those who did not report having SCT ( $n = 244$ ) on their total SCD knowledge. Total RTKQ knowledge score was not different for participants with SCT ( $M = 12.71$ ) than for those without SCT ( $M = 12.12$ ),  $t(15.3) = -1.06, p = .31$ .

Because accuracy of self-reported trait status could not be verified and many participants reported that they “thought” they had or did not have SCT, the third hypothesis was also examined in an additional manner. Participants’ responses to “Do you have sickle cell trait?” were grouped into three groups. Respondents who answered “Yes” or “I think I do” were placed into a group. Respondents who answered “I don’t know” were placed into a second group. Respondents who answered “No” or “I don’t think I do” were placed into a third group. A general linear modeling procedure was conducted to examine the contribution of SCT group status to SCKQ score. The variable of “family member with SCD” was also included in the model, as having a family member with SCD was expected to be a covariate of overall sickle cell knowledge. The overall model was significant,  $F(3,254) = 5.64, p < .001$ . The main effect of trait group was statistically significant,  $F(2,254) = 6.90, p = .001$ . Trait knowledge varied in the expected fashion. Participants who indicated that they had SCT (i.e., “Yes”/“I think I do”)

displayed the highest average SCKTKQ score ( $M = 8.20$ ). Participants who reported not having SCT (i.e., “No”/“I don’t think I do”) displayed the lowest average SCKTKQ score ( $M = 6.56$ ). The average SCKTKQ score of those who were uncertain (i.e., “I don’t know”) fell in between those of the other two groups ( $M = 7.38$ ). Having a family member with SCD did not contribute significantly to overall trait knowledge,  $F(1, 254) = 1.06, p = .31$ .

#### **Hypothesis #4**

As hypothesized, participants who reported having received sickle cell knowledge from their family (30%,  $n = 78$ ) differed significantly from participants who did not receive sickle cell knowledge from their family (70%,  $n = 180$ ) on their total sickle cell trait knowledge. A separate variances  $t$ -test showed that total SCKTKQ score was significantly higher for participants who had received information from family members ( $M = 7.42$ ) than for those who had not received information from family members ( $M = 6.53$ ),  $t(256) = -3.26, p = .001$ .

#### **Exploratory Analyses**

**Sex Differences.** Males and females differed in their overall knowledge of SCT and SCD. Females displayed significantly higher scores on overall trait knowledge ( $M = 7.09$ ) than males ( $M = 6.20$ ),  $t(256) = -3.32, p = .001$ . Females ( $M = 12.67$ ) also scored significantly higher than males ( $M = 11.08$ ) on overall disease knowledge,  $t(256) = -5.09, p < .0001$ . Fisher’s exact test was used to examine sex differences for several additional variables. Males and females responded significantly differently to the question, “Do you have sickle cell trait?”,  $\chi^2(4) = 12.16, p = .016$ . A Tukey-style multiple comparison test, which takes into account the sample size for each set of comparisons, was used to determine which pairs of responses (“No,” “I don’t think I do,” “I don’t know,” “I think I do,” and “Yes”) were significantly different (Zar, 1999). A significant difference was found between responses for “No” and “I don’t think I do” ( $q = 4.43, p < .05$ ). Females expressed more uncertainty regarding their trait status: 43% of females

endorsed “I don’t think I do” compared to 24% of males. More males reported that they did not have SCT than females (56% vs. 35%). Otherwise, the proportion of males and females endorsing each of the other three choices were similar: 1) “Yes” (5% vs. 6%), 2) “I think I do” (1% vs. 3%) and 3) “I don’t know” (14% vs. 14%). In terms of self-reported disease status, no sex differences were found,  $\chi^2(3) = 2.02, p = .57$ .

Another variable of interest in terms of possible sex differences was in the reporting of the trait status of family members. A significant between-groups difference between males and females was found in the pattern of responses for the question, “Does anyone in your family have sickle cell trait?”,  $\chi^2(2) = 8.09, p = .018$ . A Tukey-style multiple comparison test indicated that significant differences existed between the responses for “No” and “I don’t know” ( $q = 3.33, p < .05$ ), as well as between the responses for “No” and “Yes” ( $q = 3.55, p < .05$ ). More females (65%) again indicated that they were uncertain if any family members had trait than males (57%). More males (32%) stated that no family members had trait than females (17%). Additionally, more females than males indicated that a family member did have trait (18% vs. 11%). In terms of reporting the disease status of family, sex differences did not reach significance,  $\chi^2(2) = 5.03, p = .08$ .

Fisher’s exact test also indicated a significant difference between males and females in their desire to be notified of their SCT status. A much larger proportion of females (64%) requested to be notified of their SCT status than males (41%),  $\chi^2(1) = 12.08, p = .0005$ . A significant sex difference was also found in terms of who requested a summary of research findings. Again, a much greater percentage of females (53%) desired to receive a summary of research findings following the conclusion of the study than males (38%),  $\chi^2(1) = 4.93, p = .026$ .

**Predictors of trait, disease and overall sickle cell knowledge.** As in hypothesis 3, general linear modeling was also used to examine the contribution of self-reported SCT status to SCD knowledge, as measured by the RTKQ. Participants were again collapsed into three groups based upon their reported trait statuses (i.e., “Yes”/“I think I do,” “I don’t know,” “No”/“I don’t think I do”). The variable of “family member with SCD” was also included in the model, as having a family member with SCD had been hypothesized to be a covariate. The model predicting disease knowledge (RTKQ score) was not significant,  $F(3, 254) = 2.00, p = .11$ . Neither sickle cell trait group status,  $F(2,254) = 2.12, p = .12$ , nor the variable of “family member with SCD,”  $F(1,254) = .84, p = .36$ , had significant main effects. Though group trait status did not have a significant main effect on the model, disease knowledge did vary in the expected direction, with participants who had trait (“Yes”/“I think I do”) displaying the highest average RTKQ score ( $M = 13.10$ ). Participants without trait (“No”/“I don’t think I do”) displayed the lowest average RTKQ score ( $M = 11.98$ ), and those who were unsure of trait status (“I don’t know”) displayed an average RTKQ score which fell in the middle of the groups ( $M = 12.58$ ).

Multiple linear regression was used to assess a model for predicting trait knowledge (i.e., SCKQ score) using six variables: sex, trait status, disease status, total number of knowledge sources, family members with SCT, and family members with SCD. The variable of family member with SCT and the variable of family member with SCD were treated as dichotomous variables, with responses of “No” and “I don’t know” grouped together. The variables of trait status and disease status each contained five levels (i.e., “No,” “I don’t think so,” “I don’t know,” “I think so,” and “Yes”). Basic descriptive statistics and correlations for variables included in all regression models are shown in Table 4, and regression statistics are shown in Table 5. Four of

the six predictor variables—sex, trait status, family members with trait, and number of knowledge sources—had significant partial effects in the full model ( $p < .05$ ). The overall model was able to account for 17% of the variance in SCKTKQ scores,  $F(6,251) = 8.59, p < .0001$ .

Multiple linear regression was used to assess a model for predicting disease knowledge (i.e., RTKQ score) using the same variables as in the trait knowledge model—from sex, self-reported trait status, self-reported disease status, total number of sources of sickle cell knowledge, family members with SCT, and family members with SCD. Sex and total number of knowledge sources had significant partial effects ( $p < .05$ ) in the full model. The overall model accounted for 13% of the variance in overall SCD knowledge,  $F(6,251) = 6.40, p < .0001$ .

Table 1

*Sickle Cell Trait and Disease Status Descriptive Statistics*

	Total <sup>a</sup>	Females	Males
	N=258	n=174	n=84
Have you heard of the term “sickle cell trait” before?			
Yes	91%	91%	92%
No	9%	9%	8%
Do you have sickle cell trait? <sup>b</sup>			
Yes	5%	6%	5%
I think I do	2%	3%	1%
I don’t know	14%	14%	14%
I don’t think I do	36%	43%	24%
No	42%	35%	56%
Does anyone in your family have sickle cell trait? <sup>b</sup>			
Yes	16%	18%	11%
I don’t know	62%	65%	57%
No	22%	17%	32%
Have you heard of the term “sickle cell disease” before?			
Yes	97%	97%	96%
No	3%	3%	4%
Do you have sickle cell disease?			
Yes	2%	1%	4%
I think I do	0%	0%	0%
I don’t know	3%	3%	2%
I don’t think I do	18%	18%	17%
No	77%	77%	77%
Does anyone in your family have sickle cell disease?			
Yes	10%	11%	8%
I don’t know	36%	31%	45%
No	54%	57%	46%

Note. <sup>a</sup> Percentages do not always add up to 100% due to rounding.

<sup>b</sup> p < .05 between males and females

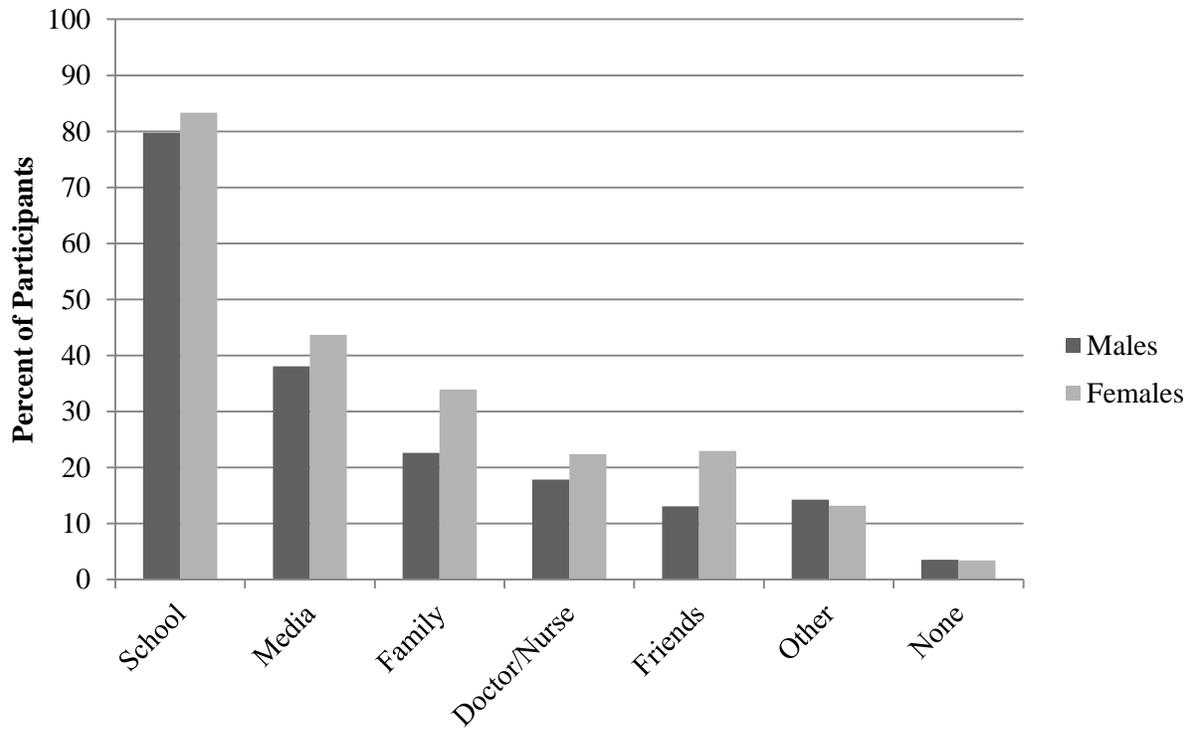


Figure 1. Percentage of male and female participants endorsing each source of knowledge.

Table 2

*Sickle Cell Trait Knowledge Questionnaire Responses (N = 266)*

	%
	Correct
1 Sickle cell trait can turn into sickle cell disease (false).	45.0%
2 If a person has sickle cell trait, it can be passed on to their children (true).	95.4%
3 Sickle cell trait causes lots of pain crises (false).	62.8%
4 People with sickle cell trait have inherited a gene for sickle hemoglobin from one parent but not the other (true).	81.8%
5 People with sickle cell trait who are athletes such as football players may be at-risk for sudden death (true). <sup>a</sup>	47.7%
6 Most people with sickle cell trait live long, healthy lives (true).	74.4%
7 If a person has sickle cell trait, all of his or her children will have sickle cell trait (false).	74.0%
8 Sickle cell trait is only passed down through the mother (false).	76.7%
9 People from all racial/ethnic backgrounds can be affected by sickle cell trait (true).	69.8%
10 Sickle cell trait causes many medical problems for most affected individuals (false).	41.9%
11 All states in the U.S. screen newborn babies for sickle cell trait as well as other conditions (true). <sup>a</sup>	78.7%
12 Sickle cell trait results in many deaths each year (false).	58.5%

*Note.* <sup>a</sup>Excluded from final overall score for SCT knowledge.

Table 3

*Revised Transition Knowledge Questionnaire Responses (N = 266)*

	%
	Correct
1 Sickle cell disease is a condition that mainly affects: a) white blood cells, b) red blood cells*, c) platelets, d) all of the above	79.1%
2 Red blood cells in sickle cell disease cause problems because they can become: a) too large, b) too soft, c) sickle-shaped and hard*, d) round and hard	85.3%
3 Hemoglobin in red blood cells carries _____ throughout the body: a) vitamins, b) minerals, c) oxygen*, d) water	84.1%
4 People with sickle cell disease have mainly hemoglobin _____ in their red blood cells: a) A, b) B, c) S*, d) E <sup>a</sup>	36.4%
5 Sickle cell disease is: a) inherited (passed down from parent to child)*, b) contagious (can be caught like a cold), c) a bleeding problem, d) caused by poor diet	95.7%
6 Hemoglobin SC disease and sickle beta thalassemia are forms of sickle cell disease: true*/false <sup>a</sup>	79.1%
7 People with sickle cell disease have inherited two genes for sickle hemoglobin, one from each parent: true*/false	59.7%
8 Why do children with sickle cell disease take penicillin every day? a) to treat infections, b) to increase appetite, c) to prevent painful episodes, d) to decrease risk of serious infection*	56.2%
9 What special doctor takes care of patients with sickle cell disease? a) family doctor, b) hematologist*, c) orthopedist, d) gynecologist	91.9%
10 In adolescents with sickle cell disease, (select one): a) sexual drive is not normal, b) physical development might be delayed*, c) females cannot have children, d) males are not able to play school sports	74.0%
11 Adolescents with sickle cell disease may: a) be shorter than others their age, b) tire more easily, c) mature later than peers, d) all of the above*	67.8%
12 A woman with sickle cell disease can complete her pregnancy with good medical care: true*/false	86.8%
13 One of the following is <b>not</b> a complication of sickle cell disease, (select one): a) priapism, b) acute chest syndrome, c) cancer*, d) leg ulcers	61.2%
14 Adolescents with sickle cell disease who are late in developing will never catch up to others: true/false*	84.1%
15 Which of the following is true: a) A female with sickle cell disease cannot have children, b) A male with sickle cell disease cannot father a child, c) A female with sickle cell disease who becomes pregnant should have special care*, d) No one with sickle cell disease should have children	85.7%
16 In cold weather, it is good practice for a person with sickle cell disease to do the following: a) wear warm clothes*, b) stay home from school or work, c) never go outside, d) cancel doctor's appointments	86.4%
17 In sickle cell disease, exercise, such as playing sports: a) causes too much weight loss, b) always causes pain crises, c) makes anemia worse, d) is okay until the person becomes tired*	52.0%
18 One of the most common symptoms of Sickle Cell Disease is: a) high levels of insulin, b) bleeding that is difficult to stop, c) pain episodes*, d) frequent seizures	65.1%

*Note.* \* Indicates correct response.

<sup>a</sup>Excluded from final overall score for SCD knowledge.

Table 4

*Correlations Among Descriptive Variables and Knowledge Scores*

Variables	1	2	3	4	5	6	7	8
1. Sex <sup>a</sup>	–							
2. Total knowledge sources	.12	–						
3. SCT status <sup>b</sup>	.12	.05	–					
4. SCD status <sup>b</sup>	-.03	-.05	.06	–				
5. Family member with SCT <sup>c</sup>	.09	.13*	.21***	-.04	–			
6. Family member with SCD <sup>c</sup>	.05	.14*	.13*	-.10	.45***	–		
7. SCT knowledge	.20**	.19**	.22***	.04	.31***	.11	–	
8. SCD knowledge	.30***	.18**	.14*	.06	.14*	.08	.35***	–
<i>M</i>	.67	2.11	.93	.35	.16	.10	6.80	12.15
<i>SD</i>	.47	1.25	1.07	.81	.36	.31	2.05	2.45
Range	0 – 1	0 – 6	0 – 4	0 – 4	0 – 1	0 – 1	0 – 10	2 – 16

Note. SCT = Sickle cell trait; SCD = Sickle cell disease.

<sup>a</sup>Sex: 0 = male, 1 = female. <sup>b</sup>SCT/SCD status: 0 = No, 1 = I don't think I do, 2 = I don't know, 3 = I think I do, 4 = Yes. <sup>c</sup>Family members with SCT/SCD: 0 = No/I don't know, 1 = Yes.

\* $p < .05$ . \*\* $p < .01$ . \*\*\* $p < .001$ .

Table 5

*Multiple Linear Regression Models for Predicting Trait and Disease Knowledge*

	Model 1 Trait Knowledge			Model 2 Disease Knowledge		
	$\beta$	$sr^2$	b	$\beta$	$sr^2$	b
<i>Variables</i>						
Sex	.15*	.02	0.65	.27***	.07	1.42
Total knowledge sources	.14*	.02	0.23	.13*	.02	0.26
Sickle cell trait status	.14*	.02	0.27	.08	.01	0.18
Sickle cell disease status	.04	.00	0.13	.07	.01	0.22
Family members with SCT	.28***	.06	1.58	.08	.00	0.53
Family members with SCD	-.06	.00	-0.39	.01	.00	0.10
<i>F</i>	8.59***			6.40***		
<i>df</i>	6			6		
<i>R</i> <sup>2</sup>	.17			.13		

\* $p < .05$ , \*\* $p < .01$ , \*\*\*  $p < .001$

## Chapter IV: Discussion

### Study Summary

In the current study, African-American college students ( $N=258$ ) were surveyed regarding their SCT and SCD status; trait and disease status of family members; and sources of knowledge of sickle cell information. Participants then completed two questionnaires designed to assess their level of knowledge of SCT and SCD. The majority of participants were female and in their late teens or early twenties. A small percentage of participants (2%) reported having children, which may have provided them with additional information regarding genetic testing because newborns are screened at birth for a variety of genetic conditions. A majority of participants (52%) were uncertain of their SCT status, as measured by responses of “I don’t know,” “I think I do,” or “I don’t think I do” when asked if they had SCT. A majority of participants (62%) did not know if anyone in their family was positive for SCT.

Participants reported few sources of knowledge of sickle cell, with the majority of participants indicating that they had received information about sickle cell from one or two sources. School was the most reported source of knowledge, and media was the second most reported source. Interestingly, when offered the opportunity to be notified of their trait status, should that information be found in the NC State Public Health Database, a significant portion of the sample (44%) indicated that they did not wish to be informed about their status.

**Main Findings.** Four hypotheses were developed to examine participants’ knowledge of SCT and SCD, participants’ reporting of disease and trait status, and sources of knowledge of sickle cell. The first hypothesis, that participants would possess low levels of overall knowledge of SCT and SCD was supported. Statistical analysis indicated that the majority of participants were familiar with the term “sickle cell trait”. However, participants scored an average of only

68% on a true/false SCT knowledge questionnaire. Detailed knowledge of SCT was limited, and participants displayed poor understanding of the genetic transmission of SCT and its typically asymptomatic presentation. A majority of participants (55%) incorrectly believed that SCT can “turn into” SCD. Over a third of respondents (37%) indicated that SCT causes many pain crises for affected individuals, and over a quarter of respondents (26%) did not believe that most people with sickle cell trait live long, healthy lives. The genetic basis of sickle cell was not well-understood, with over a quarter of respondents (26%) indicating that if a person has sickle cell trait, all of his or her children would also have SCT. Nearly a third of respondents (30%) were unaware that individuals from all races/ethnic backgrounds can be affected by SCT. A majority of participants (58%) incorrectly indicated that SCT causes many medical problems for affected individuals, and a substantial portion (41%) also believed that SCT results in many deaths each year.

Participants displayed a higher level of knowledge about SCD, though measures for the two conditions differed in a number of significant ways (i.e., number of questions, type of questions). Statistical analysis indicated that the majority of participants were familiar with the term “sickle cell disease.” However, detailed knowledge of SCD, including genetic transmission, common symptoms, prognosis, and treatment was low. No participants correctly answered all 18 questions, including the five participants who reported having SCD. Participants’ average score on the SCD knowledge questionnaire was 76%. Individual item analysis indicated that the genetic transmission of SCD was not well understood by participants. Approximately 40% were unaware that individuals with SCD inherit two genes for sickle hemoglobin, one from the mother and one from the father. A significant portion of participants (32%) were unfamiliar with common sickle cell symptoms including delayed physical

development, delayed sexual maturity, and fatigue. Complications of SCD including priapism, acute chest syndrome, and leg ulcers were identified by 39% of the sample. Over a third of participants (35%) were unable to recognize pain episodes as the most common symptom of SCD. Nearly half of participants (48%) were unaware that individuals with SCD may be able to exercise or play sports safely, and many (44%) did not know that children with sickle cell disease take penicillin daily to decrease risk of serious infection. A majority of participants (64%) were unable to identify the type of hemoglobin (i.e., S) possessed by individuals with SCD.

The second hypothesis consisted of two parts: a) that a significant number of participants would be uncertain of their sickle cell trait status and b) that there would be a significant number of false negatives or individuals who believe they do not have SCT when in fact they do. The first part of the second hypothesis was supported, but the second part was unable to be tested. The majority of respondents indicated that they were not certain of their SCT status. The NC State Laboratory of Public Health's database was accessed to determine the trait status of participants born in NC. However, newborn screening results were unable to be accessed as originally planned.

This highlights a significant problem in NC's current sickle cell reporting system. While thousands of individuals have previously been screened for SCD, thus identifying carriers of the sickle gene, results for many individuals now appear to be inaccessible, at a time when increasing evidence indicates that carrier status has some very significant, though rare, complications. Barriers to accessing past results (i.e., cumbersome search process, must request transfer of records from entity which originally requested screening, etc.) also means that trait

screenings will now be duplicated for many individuals, as college athletes are now being tested for trait prior to participation in NCAA athletic programs.

Because trait status of participants could not be verified, the third hypothesis was untestable as written. The intent of the third hypothesis was to compare the knowledge level of participants who were aware of their positive SCT status (i.e., “true positives”) to the knowledge of participants who were unaware of their positive SCT status (i.e., “false negatives”), with the variable “family member with SCD” serving as a covariate. It was hypothesized that accurate knowledge of positive SCT status would be associated with increased SCT knowledge. Having a family member with SCD was hypothesized to act as a covariate on total trait knowledge. The hypothesis was examined instead by comparing knowledge levels based on self-reported trait status. The participants who indicated that they had SCT displayed significantly higher SCT knowledge than those who did not have SCT or were not certain of their status.

Because accuracy of trait status could not be verified, the third hypothesis was also examined in an additional way. Based on their reporting of SCT status, participants were collapsed into three groups: “Yes”/“I think I do,” “I don’t know,” and “No”/“I don’t think I do.” General linear modeling was then utilized to examine knowledge among the three groups, with “family member with SCD” serving as a covariate. The overall model was statistically significant, though having a family member with SCD did not have a significant effect on overall trait knowledge. Trait knowledge did have a significant main effect on overall trait knowledge; participants who reported having SCT displayed the highest average trait knowledge score. Those who reported not having SCT had the lowest average trait knowledge score, and those who were uncertain displayed a mean trait knowledge score which was in between the other two groups.

The fourth hypothesis, that participants who had received information on sickle cell from family members would demonstrate increased knowledge over those who have received information from other sources (media, friends, teachers, or health care professionals) was supported. Participants who reported having received sickle cell knowledge from their family scored significantly higher on the trait knowledge questionnaire than those who had not receive sickle cell knowledge from their family.

**Exploratory Analyses.** Between-group post hoc analyses indicated several interesting gender differences. A significantly higher proportion of females (64%) requested to be notified of their SCT status if it were listed on the database than males (41%). Many more females (53%) than males (38%) also requested to receive a summary of research findings following the conclusion of the study.

Females and males differed significantly in their self-report of trait status, their report of the trait status of family members, and whether family was a source of knowledge about sickle cell. In general, females appeared to express greater uncertainty regarding their trait status. More females (43%) than males (24%) responded “I don’t think so” when asked about their status, while more males (56%) than females (35%) reported “No.” In terms of the trait status of family members, more females than males reported that someone in their family has SCT (18% vs. 11%) or that they were uncertain of the trait status of family members (65% vs. 57%). Many more males reported definitively that no family members had SCT (32%) compared to females (17%). Interestingly, no sex differences existed for self-report of SCD status or the SCD status of family members. Finally, males and females differed in their overall understanding of trait and disease, with females scoring significantly higher than males on measures of both trait and disease knowledge.

General linear modeling was used to examine the effect of trait status on SCD knowledge. Disease knowledge did vary by SCT group in the expected direction, though the overall model was not significant. Participants who reported that they were positive for trait or thought they were positive for it had the highest mean SCD knowledge score. Participants who reported that they did not have trait or did not “think” that they had trait had the lowest mean score on the disease knowledge questionnaire. The average score of participants who responded to “Do you have sickle cell trait?” with “I don’t know” fell in between the two other groups. However, group differences failed to reach statistical significance. Having a family member with disease was included in the models as it was expected that having a family member with SCD would be strongly associated with increased knowledge. However, this was not supported, and having a family member with SCD was not predictive of disease knowledge.

Regression analyses were conducted to evaluate whether six variables—sex, self-reported SCT status, self-reported SCD status, number of knowledge sources, family members with trait, and family members with disease—were significant predictors of trait knowledge and disease knowledge. Both models were statistically significant. Sex and total number of knowledge sources were significant predictors in each model.

Four variables had significant partial effects in predicting trait knowledge: sex, total knowledge sources, family members with SCT, and personal SCT status. Interestingly, having a family member with trait was the strongest predictor of trait knowledge. Those with family members with trait displayed better understanding of trait. Sex was the next strongest predictor, with females displaying higher levels of trait knowledge males. A greater number of knowledge sources predicted higher trait knowledge, and those who reported having trait also displayed higher SCT knowledge scores.

Two variables had significant partial effects in predicting disease knowledge: sex and total number of knowledge sources. Sex was the strongest predictor in disease knowledge, with females displaying higher levels of SCD knowledge. Again, more knowledge sources predicted greater disease knowledge.

### **Study Findings in the Context of the Literature**

**Hypothesis #1 – Knowledge of SCT and SCD.** That participants possessed low levels of understanding of SCT and SCD is consistent with previous research, though limited, which has examined knowledge of sickle cell. The current study found that while participants recognized the terms “sickle cell trait” and “sickle cell disease,” many had difficulty answering questions involving the genetic inheritance, common symptoms, prognosis, and treatment of these conditions. Only two groups of researchers have published studies assessing knowledge of sickle cell among US populations. One surveyed parents with SCT and parents of children with SCT or SCD in Chicago, and found that significant misunderstanding existed among this targeted group in terms of what it means to have SCT, as well as its health and reproductive implications (Acharya et al., 2009). The other study surveyed African-American college students in Texas regarding critical aspects of SCD and found that the basic facts of SCD were not well understood (Ogamdi, 1994).

Though Acharya et al. (2009) examined SCT knowledge, their study surveyed only a targeted sample of parents already very familiar with the condition. In contrast, the current study is the first to have assessed knowledge of trait among a large sample of African-American college students. Acharya et al. surveyed a targeted sample of parents in Chicago who either had SCT or had a child with SCT or SCD, and found significant misunderstanding of the condition. In the Chicago sample, 89% of participants understood that SCD is a genetic condition. In the

current sample, 95% of participants understood that if a person has SCT, it can be passed on to their children. While 77% of the Chicago sample understood that SCT cannot “turn into” SCD, only 45% of the current sample did. Other questions differed in wording though significant portions of both samples had difficulty answering more complex questions about inheritance of SCT (e.g., “If a person has SCT, all of his or her children will have SCT,” “Sickle cell disease can be inherited if one parent has the trait and the other parent does not”). Clearly, there is a need for further research into the level of knowledge both among targeted groups who have SCT as well as among the general population of African Americans. No studies have thus utilized the same SCT scale to directly compare SCT knowledge among different populations. Research aimed at validating a general knowledge SCT scale is also warranted.

As in the current study, Ogamdi (1994) used a multiple-choice questionnaire to assess for knowledge of SCD among 334 African-American college students in Texas. The questionnaire used by Ogamdi (1994) was created for that study and is currently unpublished. The specific wording of questions designed to assess SCD knowledge and the questionnaire’s psychometric properties are unknown. Thus comparing responses to specific content items should be done cautiously, and differences in measures likely account for some of the variance. It is of note that participants in the current sample performed better in many areas of SCD knowledge. In the current study, 79% of participants were able to correctly identify the cell that is sickled, compared with 55% of the Texas sample. When asked how SCD is transmitted, 96% of the current sample identified the correct response as “inherited (passed down from parent to child),” whereas 72% of the Texas sample correctly identified how the disease is transmitted. Approximately 87% of the current sample was able to correctly identify that women with SCD who become pregnant should have special care, though in the Texas sample 52% recognized that

pregnancy for females with SCD is a high-risk endeavor and must be carefully monitored. When asked about common symptoms of SCD, more than 65% of the current sample was able to identify pain episodes as one of the most common symptoms; Ogamdi (1994) reported that 63% of his sample was able to correctly identify characteristic symptoms of SCD. Whether SCD knowledge is increasing among African-American college students or whether regional differences may account for differences in sickle cell knowledge may be appropriate topics for future studies. The creation of standardized and validated measures for assessing trait and disease would be helpful in controlling for differences in sample to the maximum extent possible.

**Hypotheses 2a & 2b – Reporting and Accuracy of SCT Status.** No previous studies have surveyed a large group of African Americans to assess self-report of trait status and the accuracy of that report. Ogamdi (1994) indicated that “many” of the 334 African-American college students he surveyed could not recall whether they had previously been tested for SCT but provided no specific data; participants in the Ogamdi study were not asked to disclose their trait status. Of the 53 parents who had SCT or a child with SCT/SCD in the study by Acharya and colleagues (2009), three were unaware of their trait status. The majority of respondents reported becoming aware of their trait status through a pregnancy screening or after their child screened positive for SCT. In the current sample, 52% of respondents indicated that they were not certain of their trait status, responding either “I don’t know,” “I don’t think I do,” or “I think I do.”

Participants were asked to disclose their place of birth so that trait status could be verified through the NC State Public Health Laboratory database. The vast majority of participants were born in NC following 1987, the year in which mandatory newborn screenings for sickle cell

began for all African-American infants. Thus most participants in the current study were screened for SCD and SCT at birth and yet are unable to confirm their status during early adulthood—the time of life when knowledge of one’s genetic makeup may be most critical since individuals of this age have reached sexual maturity, and many already have or will soon begin to have children of their own. It may be the case that most or all of the individuals who were not certain of their status are negative for SCT. While conveying negative trait status to the individual does not have the urgency or implications of conveying a positive trait status, it is interesting that such a large portion of the sample was uncertain about such an important and relatively common health condition (i.e., 8% of the general African-American population).

Approximately 5% of the current sample indicated that they had trait. Interestingly though, self-report of trait status varied significantly by sex, with a much larger percentage of males (56%) denying that they had trait than females (35%), and more females (43%) indicating that they did not think that they had trait than males (24%). Because SCT is not a sex-linked trait, the male-to-female ratio of SCT and SCD is 1:1. Thus there is no reason to expect that fewer males than females would have trait. Instead, differences in reporting trait status may be due to gender differences in perceived susceptibility to trait, the relative importance placed on trait status in terms of reproductive planning, stigma, or some other unidentified factor.

Unfortunately, participants’ reported trait statuses were unable to be verified through the online database of the NC State Laboratory of Public Health. Laboratory staff was unable to verify whether screening results obtained prior to the mid-1990s were uploaded to the database or where their current location may be. Staff suggested that before lab results were aggregated in an online database, an individual’s screening results were likely sent to three sources: 1) mother, 2) pediatrician, and 3) local public health department. Because participants had not given

consent to contact any of these other sources, self-reported status could not be verified. Future studies may wish to recruit a younger sample whose results could theoretically be found in the online database or to identify other methods of verifying trait status.

**Hypothesis 3 – Link between Trait Status and Trait Knowledge.** No published studies have previously assessed whether knowledge of positive trait status is linked with increased sickle cell knowledge. Though trait status could not be verified, participants who indicated that they had SCT or thought they had SCT showed higher levels of trait knowledge than those who did not know their status or who reported not having SCT. Disclosing positive trait status after screening is done with the intent of conveying knowledge about the condition to the family (i.e., thus alerting them to the potential for passing SCT onto subsequent children) and/or to the affected child. Though trait status could not be verified in the current study, it is a promising sign that individuals who reported having trait displayed higher trait knowledge, including understanding of its genetic transmission. Future studies may wish to further explore what knowledge is shared to individuals who are positive for trait and who shares this knowledge (i.e., parent, physician, counselor).

**Hypothesis 4 – Family as a Knowledge Source.** No previous studies have examined whether certain knowledge sources are predictive of higher levels of sickle cell knowledge. Of particular interest in the current study is the finding that individuals who had received information about sickle cell from families displayed significantly higher levels of trait knowledge than those who had not received sickle cell information from their families. Future studies may wish to explore sources of knowledge in greater depth and seek to identify why family may be a more salient source of sickle cell knowledge.

Only one previous published study has explored sickle cell knowledge sources in general. Acharya et al. (2009) asked their targeted population of parents with SCT or who had a child who had SCT to identify sources from which they had received information about sickle cell. Participants most commonly cited pediatricians, SCD clinic staff, employees of the Sickle Cell Disease Association of Illinois, family members, and ob/gyn providers. In contrast, the most cited knowledge sources for the current sample of undergraduate African-American students included school, media, and family. That knowledge sources differed significantly is not surprising given the differences between the samples.

### **Study Strengths**

The strengths of the current study result from the nature of the sample and the novelty of the research questions being addressed. Only one previously published study has examined knowledge of SCT (Acharya et al., 2009). However, that study investigated trait knowledge among a sample of parents who either had trait or had a child with SCT or SCD. The current study fills a significant gap in the literature—examining trait knowledge among a large sample of African Americans. Approximately 8% of African Americans carry SCT, yet no study has previously investigated the level of trait knowledge among the general African American population (Tsaras, Owusu-Ansah, Owusua-Boateng, & Amoateng-Adjepong, 2009). Because the current sample had an average age of 19 years, information gained about trait knowledge may be particularly useful since by this time many individuals have become sexually active and will begin bearing children. Although Ogamdi (1994) had previously assessed knowledge of SCD among a large scale population of African-American college students, his study failed to include questions about trait.

In addition to assessing trait knowledge, the current study expanded upon previous work by addressing several novel research questions. It is the first to examine sickle cell knowledge sources among a general population of African Americans, to address whether particular knowledge sources are associated with a greater understanding of sickle cell, and to examine whether knowing one's trait status is linked with greater sickle cell knowledge. In addition, no previous studies have examined sex differences for sickle cell knowledge or reporting of trait status. The study is the first to identify sex differences which suggest that females may have greater understanding of sickle cell than males, thus identifying African-American males as a potential source of sickle cell education campaigns. In addition, the finding that number of knowledge sources was a significant predictor of higher levels of trait and disease knowledge has implications in terms of outreach and education.

### **Study Limitations**

**Measurement Issues.** One of the primary limitations of the current study is related to the measures used to assess trait and disease knowledge. The SCTKQ, a 12-item scale, was created to assess trait knowledge including the genetic transmission of sickle cell, differences between trait and disease, current screening methods, prevalence, and important implications of trait status (i.e., typically asymptomatic, rarely associated with sudden collapse, etc.). Two questions were discarded to improve the measure's internal reliability, resulting in a 10-item scale. However, the scale's internal reliability remained low. Brief measures often have low internal reliability as measured by Chronbach's alpha, since the alpha is dependent both upon the magnitude of correlations among items, as well as the number of items in the scale (Streiner & Norman, 1989). While brief scales may have utility, particularly when used within a wider battery, the SCTKQ was used in the current study as the single measure of trait knowledge.

Thus findings related to trait knowledge should be interpreted with some caution. The scale also included only true/false questions, limiting the range of participant responses. Future studies may wish to modify and to expand the SCTKQ in order to design a lengthier scale which includes several multiple-choice items for each area of interest and which has higher internal consistency. Future studies may also wish to evaluate the questionnaire for content validity, reliability, and level of readability.

In addition, the measure used to assess disease knowledge, the RTKQ, was adapted from a questionnaire designed to assess sickle cell knowledge among adolescents with SCD prior to transitioning from a pediatric to an adult clinic (i.e., Transition Knowledge Questionnaire; Newland, Cecil, & Fithian, 2000). Due to its intended population, several items had to be eliminated or modified for use in the current study. Though two questions were ultimately discarded to improve internal consistency, the final 16-item measure had a moderately low internal consistency. Like the SCTKQ, the RTKQ was brief, and creating a lengthier SCD knowledge measure which has improved consistency would be an appropriate focus of future studies. In addition, care should be taken to design a measure with the intent of assessing knowledge among non-SCD adults, as many of the items used in the current scale were related to sickle cell care and symptoms in adolescence.

**Generalizability Issues.** Participants in the current study consisted of undergraduate African-American students from a large public university in the southeastern United States. Although this sample was ideal in many ways, it may limit the ability to generalize findings to the larger African-American population. College-educated individuals may have a higher level of knowledge of sickle cell or a better understanding of genetic transmission than those without a college education. Participants in the current sample selected “school” as the most common

source of their sickle cell knowledge. Individuals at a university may have had more opportunities to learn about sickle cell or other genetic disorders through high school or college courses. In addition to knowledge level, differences may exist between college-educated individuals and those with less education in their consumption of health information, as well as familiarity with their own medical history (i.e., trait status). Individuals with higher levels of education may also be exposed to more potential knowledge sources than the general population. While participants were not asked for any demographic information related to socio-economic status (SES), a well-established link exists between education and SES. Knowledge may vary not only across education level, but also likely across economic level. Future studies may wish also to assess for SES among participants in order to examine whether differences exist.

Also, because states vary considerably in their reporting of sickle cell knowledge and the resources available for disseminating sickle cell knowledge (Kavanagh et al., 2008), it may be the case that regional differences contribute to the level of knowledge individuals possess. Future studies may wish to explore whether regional differences exist in terms of sickle cell familiarity, as well as the source of any potential differences (i.e., percentage of African-American residents, variations in state sickle cell associations, etc.)

**Validity Issues.** One final limitation is related to the validity of participants' self-report of trait status. A primary purpose of the current study was to identify whether there are individuals in North Carolina who were screened at birth and found to have SCT, yet who, in early adulthood, are unaware of their positive trait status. Unfortunately, the actual trait status of participants could not be confirmed through the state laboratory's online database. Given that the majority of participants were uncertain of their status, it is probable that some individuals may have inaccurately reported their statuses. In such cases, participants may have either

reported that they did have SCT when in fact they did not, or that they did not have SCT when in fact they did. Because SCT is asymptomatic, individuals likely assume that they do not have SCT unless provided with evidence to the contrary. Thus, it is likely that there are participants who may have SCT and be unaware of their status. Several key findings from the current study were related to trait status, including that self-report of positive trait status is linked with higher levels of trait knowledge and that females and males differed in their reporting of trait status. It is important to acknowledge the potential limitation associated with inaccurate self-report of trait status.

### **Future Directions**

Determining how accurately African-American individuals are able to report their trait status remains a major focus for future research. The current study provides initial evidence that a significant portion of African Americans may be uncertain of their trait status. Future studies should seek to determine how information about SCT status can best be transmitted to affected individuals. With universal newborn screening for SCD now in place in all 50 states, every infant with SCT is now theoretically identified at birth. An accurate understanding of positive trait status is important for these individuals as they age, both to understand the potential for genetic transmission of SCT to future offspring as well as the rare but serious complications of SCT. Identifying whether knowledge from newborn screenings is accurately transmitted to the family and how ultimately this information can be communicated to affected individuals (via families, health care providers or state agencies) should be a focus of future studies.

A second area ripe for further investigation involves the development of better measures designed to assess knowledge of SCT and SCD. Few previous studies have examined what individuals know about these conditions, and thus limited time and effort have gone into

designing optimal knowledge scales. Although a SCT knowledge questionnaire was created for the current study, it consisted of just 12 questions and also utilized a true/false format.

Developing psychometrically sound measures which have been normed on large groups on non-affected individuals may be an appropriate focus of future studies. It may be informative to expand the SCT knowledge questionnaire used in the current study to include additional items and a multiple choice format, which would provide for an increased range of responses so that knowledge levels could be more accurately determined. Furthermore, researchers may wish to include additional items so that have multiple questions assess each area of SCT knowledge (i.e., genetic inheritance, newborn screening, etc).

For disease knowledge, the current study utilized a scale designed to assess knowledge among adolescents with SCD. No scale has yet been published which has the purpose of assessing awareness and knowledge of SCD among non-affected individuals, and the creation of such a scale is warranted.

Finally, a practical goal of investigations into trait knowledge is ultimately to improve the transmission of knowledge in order to allow individuals access to and understanding of their personal health information. Surprisingly, a substantial portion of participants (44%) indicated that they did not wish to be informed of their trait status. Although accurate knowledge of trait status may be a goal of medical providers, there may be stigma associated with positive trait status which caused many participants to decline to be informed of their status. Participants may also have declined to be informed of their status for other reasons. Some may not recognize the prevalence of the condition among the general population or may not know the rare but serious complications associated with the condition which make it important for individuals to know their status. Future studies should explore factors which may inhibit young African-American

adults from desiring to learn their trait status, including whether stigma is associated with this condition among the general population.

In addition, the present study provides initial evidence, though correlational, that individuals who receive sickle cell information from their families may have a better understanding of the condition than those who have learned about sickle cell from other sources. Additional large scale studies are needed to confirm this and other research findings from the current study. However, future investigations may also wish to shift the setting of research from the laboratory to the family room, in order to complete qualitative interviews with families of children with SCT. Such interviews may provide invaluable information about when and how knowledge of carrier status is transmitted to the affected individual, as well as any fears or misconceptions that parents may have regarding the condition. Studies may also wish to examine whether there are any resources available for parents to assist them with sharing trait status with their children. Collaboration with state sickle cell agencies could provide an excellent opportunity to learn how sickle cell knowledge is currently being disseminated and to develop guidelines and resources intended to improve understanding of the condition both among affected individuals as well as the general population.

## References

- Acharya, K., Walsh Lang, C., Friedman Ross, L. (2009). A pilot study to explore knowledge, attitudes, and beliefs about sickle cell trait and disease. *Journal of the National Medical Association, 101, 11*, 1163-1172.
- Adams, J. Q. (1957). Sudden death in pregnancy due to the sickle cell trait. *Southern Medical Journal, 50(7)*, 898-901.
- Adewuyi, J.O. (2000). Knowledge of an attitudes to sickle cell disease and sickle carrier screening among new graduates of Nigerian tertiary educational institutions. *Nigerian Postgrad Medical Journal, 7*, 120-123.
- Ajayi, A. A. L. (2005). Should the sickle cell trait be reclassified as a disease state? *European Journal of Medicine, 16*, 463.
- Austin, H., Lally, C., Benson, J., Whitsett, C., Hooper, W., & Key, N. (2009). Hormonal contraception, sickle cell trait, and risk for venous thromboembolism among African American women. *American Journal of Obstetrics and Gynecology, 6*, 620.e1-620.e3.
- Ballas, S. K. (2007). Current issues in sickle cell pain and its management. *Hematology, 97-105*.
- Ballas, S. K. & Lusardi, M. (2005). Hospital readmission for adult acute sickle cell painful episodes: Frequency, etiology, and prognostic significance. *American Journal of Hematology, 79*, 17-25.
- Bauer, J. (1940). Sickle cell disease. *Archives of Surgery, 41*, 1344-1362.
- Booth, C., Inusa, B., & Obaro, S. K. (2010). Infection in sickle cell disease: A review. *International Journal of Infectious Diseases, 14(1)*, e2-e1.

Boyd, J. H., Watkins, A. R., Price, C. L., Fleming, F., & DeBaun, M. R. (2005).

Inadequate community knowledge about sickle cell disease among African-American women. *Journal of the National Medical Association, 97*(1), 62-67.

Centers for Disease Control and Prevention. (2009). *YBRSS: Youth Risk Behavior and Surveillance System* [Data file]. Retrieved from <http://www.cdc.gov/healthyyouth/yrbs/index.htm>.

Committee on Fetus and Newborn. (1965). Screening of newborn infants for metabolic disease. *Pediatrics, 35*, 499-501.

Davis, C. J. Mostofi, F. K., & Sesterhenn, I. A. (1995). Renal medullary carcinoma: The seventh sickle cell nephropathy. *The American Journal of Surgical Pathology, 19*(1), 1-11.

Davis, H., Schoendorf, K. C., Gergen, P. J., & Moore, R. M. (1997). National trends in the mortality of children with sickle cell disease, 1968 through 1992. *American Journal of Public Health, 87*(8), 1317-1322.

Farrell, M. H., Certain, L. K., & Farrell, P. M. (2001). Genetic counseling and risk communication services of newborn screening programs. *Archives of Pediatric and Adolescent Medicine, 155*, 120-126.

Grover, R., Shahidi, S., Fisher, B., Goldberg, D., & Wethers, D. (1983). Current sickle cell screening program for newborns in New York City, 1979-1980. *American Journal of Public Health, 73*(3), 249-252.

Headings, V. & Fielding, J. (1975). Guidelines for counseling young adults with sickle cell trait. *American Journal of Public Health, 65*(8), 819-827.

- Herrick, J. B. (1910). Peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia. *Archives of Internal Medicine*, VI, 517-521.
- Hill, S. A. (1994). Motherhood and the obfuscation of medical knowledge: The case of sickle cell disease. *Gender & Society*, 8(1), 29-47.
- John, A. B., Ramlal, A., Jackson, H., Maude, G. H., Waight Sharma, A., & Serieant, G. R. (1984). Prevention of pneumococcal infection in children with homozygous sickle cell disease. *British Medical Journal*, 288, 1567-1570.
- Kavanagh, P. L., Wang, C. J., Therrell, B. L., Sprinz, P. G., & Bauchner, H. (2008). Communication of positive newborn screening results for sickle cell disease and sickle cell trait: Variation across states. *American Journal of Medical Genetics*, 148, 15-22.
- Kerle, K. K. & Nishimura, K. D. (1996). Exertional collapse and sudden death associated with sickle cell trait. *American Family Physician*, 54, 237-240.
- Khatib, R., Rabah, R., & Sarnaik, S. A. (2009). The spleen in the sickling disorders: An update. *Pediatric Radiology*, 39, 17-22.
- Kirkman, H. N. (2008). Newborn screening in North Carolina: The evolution of policy and practice. *North Carolina Medical Journal*, 69(2), 92-97.
- Kirylyuk, K., Jadoon, A., Gupta, M., & Radhakrishnan, J. (2007). Sickle cell trait and gross hematuria. *Kidney International*, 71, 706-710.
- Listernick, R., Firson, L., & Silverman, B. L. (1992). Delayed diagnosis of infants with abnormal neonatal screens. *Journal of the American Medical Association*, 267(8), 1095-1099.
- National Collegiate Athletic Association. (2009). NCAA recommends testing to confirm sickle cell trait status in student-athletes. Retrieved from <http://www.ncaa.org/>.

- National Athletic Trainers' Association. (2007). Consensus statement: Sickle cell trait and the athlete.
- National Institutes of Health. (1996). *Facts about sickle cell anemia* (NIH Publication No. 96-4057). Bethesda, MD: U.S. Government Printing Office.
- National Institutes of Health. (2002). *The management of sickle cell disease* (NIH Publication No. 02-2117). Bethesda, MD: U.S. Government Printing Office.
- National Newborn Screening and Genetics Resource Center. (2009). State map newborn screening. Retrieved from <http://genes-r-us.uthscsa.edu/resources/consumer/statemap.htm>.
- National Newborn Screening and Genetics Resource Center. (2009). National newborn screening status report. Retrieved from <http://genes-r-us.uthscsa.edu/nbsdisorders.htm>.
- NC Healthy Schools (2009). *Teen pregnancy prevention*. Retrieved from <http://www.nchealthyschools.org/teenpregnancy>.
- Newborn Screening Saves Lives Act of 2007, Pub. L. No. 110-204, 110 Congress, (2008).
- NC Division of Public Health State Laboratory. (2009). Newborn screening. Retrieved from <http://slph.state.nc.us/newborn/default.asp>.
- Newland, J. A., Cecil, R., & Fithian, J. (2000). *Measurement of knowledge of sickle cell disease (SCD) in adolescents in preparation for transition to adult care*. Poster presented at the 24<sup>th</sup> Annual Meeting of the National Sickle Cell Program, Sponsored by the Comprehensive Sickle Cell Center, the Children's Hospital of Philadelphia and the National Heart, Lung and Blood Institute, National Institutes of Health, Philadelphia, PA.
- Newland, J. A. (2008). Factors influencing independence in adolescents with sickle cell disease. *Journal of Child and Adolescent Psychiatric Nursing*, 21(3), 177-185.

- North Carolina Sickle Cell Syndrome Program. (2009). Resources. Retrieved from [http://www.ncsicklecellprogram.org/SC\\_Resources.htm](http://www.ncsicklecellprogram.org/SC_Resources.htm).
- Ogamdi, S. O. (1994). African American students' awareness of sickle cell disease. *Journal of the American College Health Association, 42*(5), 234-236.
- Platt, O. S., Brambilla, D. J., Rosse, W. F., Milner, P. F., Castro, O., Steinberg, M. H., & Klug, P. P. (1994). Mortality in sickle cell disease—Life expectancy and risk factors for early death. *New England Journal of Medicine, 330*, 1639-1644.
- Sheinin, L. & Wetli, C. V. (2009). Sudden death and sickle cell trait. *American Journal of Forensic Medical Pathology, 30*(2), 204-208.
- Stoddard, J. J. & Farrell, P. M. (1997). State-to-state variations in newborn screening procedures. *Archives of Pediatric and Adolescent Medicine, 151*, 561-564.
- Streiner, D.L. & Norman, G.R. (1989). *Health measurement scales: A practical guide to their development and use*. New York: Oxford Medical Publications.
- Therrell, B. L. (2001). U.S. newborn screening policy dilemmas for the twenty-first century. *Molecular Genetics and Metabolism, 74*, 64-74.
- Therrell, B., Lorey, F., Eaton, R., Frazier, D., Hoffman, G., Boyle, C., Green, D., Devine, O., & Hannon, H. (2008). Impact of expanded newborn screening—United States, 2006. *Morbidity and Mortality Weekly Report, 57*(37), 1012-1015.
- Tsaras, G., Owusu-Ansah, A., Osusua Boateng, R., & Amoateng-Adjepong, Y. (2009). Complications associated with sickle cell trait: A brief narrative review. *The American Journal of Medicine, 122*, 507-512.

- Treadwell, M. J., McClough, L., & Vichinsky, E. (2006). Using qualitative and quantitative strategies to evaluate knowledge and perceptions about sickle cell disease and sickle cell trait. *Journal of the National Medical Association, 98*(5), 704-710.
- U.S. Department of Education. Institute of Education Sciences, National Center for Education Statistics. (2009). *Common core of data* [Data file]. Retrieved from <http://nces.ed.gov/>.
- Verduzco, L. A. & Nathan, D. G. (2009). Sickle cell disease and stroke. *Blood, 114*(25), 5117-25.
- Vichinsky, E. P., Neumayr, L. D., Earles, A. N., Williams, R., Lennette, E. T., Dean, D., Nickerson, B., Orringer, E., McKie, V., Bellevue, R., Daeschner, C., & Manci, E. A. (2000). Causes and outcomes of the acute chest syndrome in sickle cell disease. *New England Journal of Medicine, 342*, 1855-1865.
- Van Camp, S. P., Bloor, C. M., Mueller, F. O., Cantu, R. C., Olson, H. G., (1995). Nontraumatic sports death in high school and college athletes. *Medical Science of Sports and Exercise, 27*, 641-647
- Watanabe, I. C., Billis, A., Guimaraes, M. S., Alvarenga, M., de Matos, A., Cardinalli, I. A., Filippi, R. Z., de Castro, M. G., & Suzigan, S. (2007). Renal medullary carcinoma: Report of seven cases from Brazil. *Modern Pathology, 20*, 914-920.
- World Health Organization. (2006). Sickle cell anemia: Report by the secretariat (WHO Publication No. A59/9). Geneva, Switzerland: World Health Assembly.
- Zar, J.H. (1999). *Biostatistical Analysis*. Upper Saddle River, New Jersey: Pearson Prentice Hall.

## **Appendix A: Demographic Questionnaire**

## **A Little About YOU!**

1. What is your **name**?

\_\_\_\_\_

First

Middle

Last

2. Is this the name that was given to you **when you were born**?

\_\_\_\_ Yes

\_\_\_\_ No

3. If it was not, what was the name given to you at birth?

\_\_\_\_\_

First

Middle

Last

4. **When** were you born? \_\_\_\_\_ / \_\_\_\_\_ / \_\_\_\_\_

Month

Day

Year

5. **Where** were you born? \_\_\_\_\_, \_\_\_\_\_

City

State

6. What is your **mother's** maiden name (the name she was born with)?

\_\_\_\_\_

First

Middle

Last (Maiden)

7. Are you: \_\_\_\_\_ MALE  
\_\_\_\_\_ FEMALE  
(please mark or circle)

8. What is your ethnic background? \_\_\_\_\_ African-American  
\_\_\_\_\_ Caucasian  
\_\_\_\_\_ Hispanic  
\_\_\_\_\_ Biracial  
\_\_\_\_\_ Other  
(please mark or circle)

9. Are you a parent? \_\_\_\_\_ YES  
\_\_\_\_\_ NO

## **Appendix B: Personal and Family Health Questionnaire**

## **Personal & Family Health Questionnaire**

Please check your responses to the following questions.

1. Have you heard of the term “**sickle cell disease**” before?
  - Yes
  - No
  
2. Do you have **sickle cell disease**?
  - Yes
  - I think I do
  - I don't know
  - I don't think I do
  - No
  
3. Does anyone in your family have **sickle cell disease**?
  - Yes → Please check: Mother Father Sister Brother Aunt/Uncle Daughter  
Cousin Grandmother/Grandfather Son Other
  - No
  - I don't know
  
4. Have you heard of the term “**sickle cell trait**” before?
  - Yes
  - No
  
5. Do you have **sickle cell trait**?
  - Yes
  - I think I do
  - I don't know
  - I don't think I do
  - No
  
6. Does anyone in your family have **sickle cell trait**?
  - Yes→ Please check: Mother Father Sister Brother Aunt/Uncle Daughter  
Cousin Grandmother/Grandfather Son Other
  - No
  - I don't know
  
7. Where you have learned about sickle cell. (Check **all** that apply)
  - At school
  - From family
  - From friends
  - From a doctor or nurse
  - From the TV, movies, or something you read
  - Other \_\_\_\_\_
  - I have **not** received any information about sickle cell

## **Appendix C: Sickle Cell Trait Knowledge Questionnaire**

## Sickle Cell Trait Knowledge Questionnaire

DIRECTIONS: This is a questionnaire about **SICKLE CELL TRAIT**. Circle the best answer.

1. Sickle cell trait can turn into Sickle Cell Disease.
  - A. True
  - B. False
  
2. If a person has sickle cell trait, it can be passed on to their children.
  - A. True
  - B. False
  
3. Sickle cell trait causes lots of pain crises.
  - A. True
  - B. False
  
4. People with sickle cell trait have inherited a gene for sickle hemoglobin from one parent but not the other.
  - A. True
  - B. False
  
5. People with sickle cell trait who are athletes such as football players may be at-risk for sudden death.
  - A. True
  - B. False
  
6. Most people with sickle cell trait live long, healthy lives.
  - A. True
  - B. False
  
7. If a person has sickle cell trait, all of his or her children will have sickle cell trait.
  - A. True
  - B. False
  
8. Sickle cell trait is only passed down through the mother.
  - A. True
  - B. False
  
9. People from all racial/ethnic backgrounds can be affected by sickle cell trait.
  - A. True
  - B. False
  
10. Sickle cell trait causes many medical problems for most affected individuals.
  - A. True
  - B. False

11. All states in the U.S. screen newborn babies for sickle cell trait as well as other conditions.
  - A. True
  - B. False
  
12. Sickle cell trait results in many deaths each year.
  - A. True
  - B. False

## **Appendix D: Transition Knowledge Questionnaire**

COMPREHENSIVE SICKLE CELL CENTER  
Division of Hematology  
The Children's Hospital of Philadelphia  
34<sup>th</sup> Street and Civic Center Boulevard, Philadelphia, PA 19104. (215) 590-3423

TRANSITION KNOWLEDGE QUESTIONNAIRE

Name or ID # \_\_\_\_\_

DIRECTIONS: Circle the best answer.

1. Sickle cell disease is a condition that mainly affects
  - A. white blood cells
  - B. red blood cells
  - C. platelets
  - D. all of the above
  
2. Red blood cells in sickle cell disease cause problems because they can become
  - A. too large
  - B. too soft
  - C. sickle-shaped and hard
  - D. round and hard
  
3. Hemoglobin in red blood cells carries \_\_\_\_\_ throughout the body.
  - A. Vitamins
  - B. minerals
  - C. oxygen
  - D. water
  
4. People with sickle cell disease have mainly hemoglobin \_\_\_\_\_ in their red blood cells.
  - A. A
  - B. B
  - C. S
  - D. E
  
5. Sickle cell disease is
  - A. inherited (passed down from parent to child)
  - B. contagious (can be caught like a cold)
  - C. a bleeding problem
  - D. caused by a poor diet

6. Hemoglobin SC disease and sickle beta thalassemia are forms of sickle cell disease.
- A. True
  - B. False
7. When having sickle cell pain, a patient should
- A. drink plenty of fluids
  - B. always stay home from work or school
  - C. go to the Emergency Room right away
  - D. limit intake of food
8. People with sickle cell trait have inherited two genes for sickle hemoglobin, one from each parent.
- A. True
  - B. False
9. Why do children with sickle cell disease take penicillin every day?
- A. to treat infections
  - B. to increase appetite
  - C. to prevent painful episodes
  - D. to decrease risk of serious infection
10. It is important to keep medical appointments
- A. only when I am sick
  - B. if I need stronger pain medication
  - C. even when I am having no problems
  - D. only after being discharged from the hospital
11. What special doctor takes care of patients with sickle cell disease?
- A. family doctor
  - B. hematologist
  - C. orthopedist
  - D. gynecologist
12. In adolescents with sickle cell disease, (select one)
- A. sexual drive is not normal.
  - B. physical development might be delayed.
  - C. females cannot have children.
  - D. males are not able to play school sports.

13. Adolescents with sickle cell disease may
- A. be shorter than others their age
  - B. tire more easily
  - C. mature later than peers
  - D. all of the above
14. A woman with sickle cell disease can complete her pregnancy with good medical care.
- A. True
  - B. False
15. One of the following is **not** a complication of sickle cell disease. (select one)
- A. priapism
  - B. acute chest syndrome
  - C. cancer
  - D. leg ulcers
16. It is important to keep doctors' appointments to
- A. identify any new problems with sickle cell disease
  - B. have blood tests (for example, to check blood count)
  - C. check on growth and development
  - D. all of the above
17. Adolescents with sickle cell disease who are late in developing will never catch up to others.
- A. True
  - B. False
18. Which of the following is true?
- A. A female with sickle cell disease cannot have children.
  - B. A male with sickle cell disease cannot father a child.
  - C. A female with sickle cell disease who becomes pregnant should have special care.
  - D. No one with sickle cell disease should have children.
19. Sickle cell pain is **best** treated
- A. after the pain is severe
  - B. at home when symptoms first begin
  - C. in the hospital
  - D. with narcotics only

20. Before going to the Emergency Room, a patient should first try to notify his/her regular doctor.
- A. True
  - B. False
21. A hospital Emergency Room is for emergency care only. Which is a good reason to go to the ER? (select one)
- A. pain that does not get better 2 hours after taking Tylenol
  - B. missed clinic appointment today
  - C. chest pain and trouble breathing
  - D. stuffy nose (head cold).
22. In cold weather, it is good practice for a person with sickle cell disease to do the following.
- A. Wear warm clothes.
  - B. Stay home from school or work.
  - C. Never go outside.
  - D. Cancel doctors' appointments.
23. In sickle cell disease, exercise, such as playing sports
- A. causes too much weight loss
  - B. always causes pain crises.
  - C. makes anemia worse.
  - D. is okay until the person becomes tired.
24. Hydroxyurea is a
- A. chemical found in the urine
  - B. medication used to treat sickle cell disease
  - C. narcotic
  - D. part of normal red blood cells
25. An adolescent with sickle cell disease is trying to decide where to go to college. What things must be considered?
- A. how near it is to medical care
  - B. ability to manage small health problems alone
  - C. rules of the school regarding absences due of illness
  - D. all of the above

## **Appendix E: Revised Transition Knowledge Questionnaire**

## REVISED TRANSITION KNOWLEDGE QUESTIONNAIRE

DIRECTIONS: This is a questionnaire about **SICKLE CELL DISEASE**. Circle the best answer.

1. Sickle cell disease is a condition that mainly affects
  - A. white blood cells
  - B. red blood cells
  - C. platelets
  - D. all of the above
  
2. Red blood cells in sickle cell disease cause problems because they can become
  - A. too large
  - B. too soft
  - C. sickle-shaped and hard
  - D. round and hard
  
3. Hemoglobin in red blood cells carries \_\_\_\_\_ throughout the body.
  - A. Vitamins
  - B. minerals
  - C. oxygen
  - D. water
  
4. People with sickle cell disease have mainly hemoglobin \_\_\_\_\_ in their red blood cells.
  - A. A
  - B. B
  - C. S
  - D. E
  
5. Sickle cell disease is
  - A. inherited (passed down from parent to child)
  - B. contagious (can be caught like a cold)
  - C. a bleeding problem
  - D. caused by a poor diet
  
6. Hemoglobin SC disease and sickle beta thalassemia are forms of sickle cell disease.
  - A. True
  - B. False

7. People with sickle cell disease have inherited two genes for sickle hemoglobin, one from each parent.
- A. True
  - B. False
8. Why do children with sickle cell disease take penicillin every day?
- A. to treat infections
  - B. to increase appetite
  - C. to prevent painful episodes
  - D. to decrease risk of serious infection
9. What special doctor takes care of patients with sickle cell disease?
- A. family doctor
  - B. hematologist
  - C. orthopedist
  - D. gynecologist
10. In adolescents with sickle cell disease, (select one)
- A. sexual drive is not normal.
  - B. physical development might be delayed.
  - C. females cannot have children.
  - D. males are not able to play school sports.
11. Adolescents with sickle cell disease may
- A. be shorter than others their age
  - B. tire more easily
  - C. mature later than peers
  - D. all of the above
12. A woman with sickle cell disease can complete her pregnancy with good medical care.
- A. True
  - B. False
13. One of the following is **not** a complication of sickle cell disease. (select one)
- A. priapism
  - B. acute chest syndrome
  - C. cancer
  - D. leg ulcers

14. Adolescents with sickle cell disease who are late in developing will never catch up to others.
- A. True
  - B. False
15. Which of the following is true?
- A. A female with sickle cell disease cannot have children.
  - B. A male with sickle cell disease cannot father a child.
  - C. A female with sickle cell disease who becomes pregnant should have special care.
  - D. No one with sickle cell disease should have children.
16. In cold weather, it is good practice for a person with sickle cell disease to do the following.
- A. Wear warm clothes.
  - B. Stay home from school or work.
  - C. Never go outside.
  - D. Cancel doctors' appointments.
17. In sickle cell disease, exercise, such as playing sports
- A. causes too much weight loss
  - B. always causes pain crises.
  - C. makes anemia worse.
  - D. is okay until the person becomes tired.
18. One of the most common symptoms of Sickle Cell Disease is
- A. High levels of insulin
  - B. Bleeding that is difficult to stop
  - C. Pain episodes
  - D. Frequent seizures

## **Appendix F: IRB Documentation**



**University and Medical Center Institutional Review Board**  
East Carolina University, 600 Moye Boulevard  
1L-09 Brody Medical Sciences Bldg. • Greenville, NC 27834  
Office 252-744-2914 • Fax 252-744-2284 • [www.ecu.edu/irb](http://www.ecu.edu/irb)  
Chair and Director of Biomedical IRB: L. Wiley Nifong, MD  
Chair and Director of Behavioral and Social Science IRB: Susan L. McCammon, PhD

---

TO: Tamara Warren, PhD, Dept of Psychology, ECU—104 Rawl Building, Mailstop 565  
FROM: UMCIRB *kk*  
DATE: April 8, 2010  
RE: Expedited Category Research Study  
TITLE: “Knowledge of Sickle Cell Disease and Trait Status Among African-American College Students”

**UMCIRB #10-0153**

This research study has undergone review and approval using expedited review on 4.6.10. This research study is eligible for review under an expedited category number 5 & 7. The Chairperson (or designee) deemed this **unfunded** study **no more than minimal risk** requiring a continuing review in **12 months**. Changes to this approved research may not be initiated without UMCIRB review except when necessary to eliminate an apparent immediate hazard to the participant. All unanticipated problems involving risks to participants and others must be promptly reported to the UMCIRB. The investigator must submit a continuing review/closure application to the UMCIRB prior to the date of study expiration. The investigator must adhere to all reporting requirements for this study.

The above referenced research study has been given approval for the period of **4.6.10 to 4.5.11**. The approval includes the following items:

- Internal Processing Form (dated 3.3.10)
- Debriefing
- Informed Consent (dated 4.6.10)
- Sickle Cell Disease Knowledge Questionnaire (received 3.8.10)
- Sickle Cell Trait Knowledge Questionnaire (received 3.8.10)
- Personal and Family Health Questionnaire (received 3.8.10)
- A Little About You (received 3.8.10)

The Chairperson (or designee) does not have a potential for conflict of interest on this study.

**The UMCIRB applies 45 CFR 46, Subparts A-D, to all research reviewed by the UMCIRB regardless of the funding source. 21 CFR 50 and 21 CFR 56 are applied to all research studies under the Food and Drug Administration regulation. The UMCIRB follows applicable International Conference on Harmonisation Good Clinical Practice guidelines.**

UMCIRB HIPAA Authorization Checklist/Approval Form

UMCIRB #: 10-0153 PI: Tamara Warner, PhD  
Title of study (full or abbreviated): "Knowledge of Sickle Cell Disease and Trait- Status Among African - American College Students."

Check one of the boxes below:

- Use of ECU "Research Participant Authorization to Use and Disclose Information for Research"
Use of a sponsor/granting agency or other alternative HIPAA Patient Authorization
Use of research informed consent document form with required elements of the HIPAA Patient Authorization

Designated UMCIRB reviewer has reviewed the substitute HIPAA Patient Authorization for Research or proposed research consent form and found that it is written in plain language and contains:

- Yes No
A specific and meaningful description of the information to be used or disclosed
The name or identification of persons or class of persons authorized to make requested use/disclosure of PHI
The name or identification of persons or class of persons who will use PHI for research-related purposes
A description of each purpose of the use or disclosure
The individual's signature (or that of his/her authorized representative) and the date.
An expiration date or event, or a statement "end of research study" or "none" when appropriate
A statement that the individual may revoke the authorization in writing;
Any exceptions to the right to revoke (e.g. researcher may continue to use and disclose, for research integrity and reporting purposes any PHI collected from the individual pursuant to such Authorization before it was revoked).
A statement that information disclosed under the Authorization could potentially be re-disclosed by the recipient and would no longer be protected under HIPAA.
A statement of the ability or inability to condition treatment, payment, enrollment or eligibility for benefits on the authorization by stating either stating the applicable conditions or the consequences to the individual for refusal to sign the authorization.
All the above elements are present, HIPAA AUTHORIZATION document is APPROVED
All the above elements are not present; HIPAA AUTHORIZATION document is NOT APPROVED

Designated UMCIRB Reviewer

4/12/2010
Date

Principal Investigator: Present this signed form at the time PHI is requested from custodians of records. By signing this document, I acknowledge and affirm that all enrolled subjects have signed a valid HIPAA Authorization Form.

Principal Investigator

Date



# EAST CAROLINA UNIVERSITY

University & Medical Center Institutional Review Board Office  
1L-09 Brody Medical Sciences Building • 600 Moye Boulevard • Greenville, NC 27834  
Office 252-744-2914 • Fax 252-744-2284 • [www.ecu.edu/irb](http://www.ecu.edu/irb)

TO: Tamara Warner, PhD, Dept. of Psychology, ECU—104 Rawl Building, Mailstop: 565

FROM: UMCIRB *KK*

DATE: March 22, 2011

RE: Expedited Continuing Review of a Research Study

TITLE: "Knowledge of Sickle Cell Disease and Trait Status Among African-American College Students"

## UMCIRB #10-0153

The above referenced research study was initially reviewed and approved by expedited review on 4.6.10. This research study has undergone a subsequent continuing review using expedited review on 3.21.11. This research study is eligible for expedited review because it is a research involving materials (data, documents, records, or specimens) that have been collected, or will be collected solely for nonresearch purposes (such as medical treatment or diagnosis). (NOTE: Some research in this category may be exempt from the HHS regulations for the protection of human subjects. 45 CFR 46.101(b)(4). This listing refers only to research that is not exempt.) It is also a research on individual or group characteristics or behavior (including, but not limited to, research on perception, cognition, motivation, identity, language, communication, cultural beliefs or practices, and social behavior) or research employing survey, interview, oral history, focus group, program evaluation, human factors evaluation, or quality assurance methodologies. (NOTE: Some research in this category may be exempt from the HHS regulations for the protection of human subjects. 45 CFR 46.101(b)(2) and (b)(3). This listing refers only to research that is not exempt.)

The Chairperson (or designee) deemed this **unfunded** study **no more than minimal risk** requiring a continuing review in **12 months**. Changes to this approved research may not be initiated without UMCIRB review except when necessary to eliminate an apparent immediate hazard to the participant. All unanticipated problems involving risks to participants and others must be promptly reported to the UMCIRB. The investigator must submit a continuing review/closure application to the UMCIRB prior to the date of study expiration. The investigator must adhere to all reporting requirements for this study.

The above referenced research study has been given approval for the period of **3.21.11 to 3.20.12**. The approval includes the following items:

- Continuing Review Form (date 3.11.11)
- Protocol Summary
- Informed consent (version date 4.6.10)
- Debriefing
- A Little About You
- Personal & Family Health Questionnaire
- Sickle Cell Knowledge Questionnaire
- Sickle Cell Trait Knowledge Questionnaire

The Chairperson (or designee) does not have a conflict of interest on this study.

**The UMCIRB applies 45 CFR 46, Subparts A-D, to all research reviewed by the UMCIRB regardless of the funding source. 21 CFR 50 and 21 CFR 56 are applied to all research studies under the Food and Drug Administration regulation. The UMCIRB follows applicable International Conference on Harmonisation Good Clinical Practice guidelines.**

## **Appendix G: Informed Consent**

**Informed Consent to Participate in Research &  
Research Participant Authorization to Use and Disclose Protected Health Information**  
Information to consider before taking part in research that has no more than minimal risk.

**Title of Research Study: Knowledge of Sickle Cell Disease and Trait Status Among African-American College Students**

Principal Investigator: Tamara D. Warner, Ph.D.  
Institution/Department or Division: East Carolina University, Department of Psychology  
Address: 104 Rawl Building  
Telephone #: (252) 328-6282

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Researchers at East Carolina University (ECU) study problems in society, health problems, environmental problems, behavior problems and the human condition. Our goal is to try to find ways to improve the lives of you and others. To do this, we need the help of people who are willing to take part in research.

The person who is in charge of this research is called the Principal Investigator. The Principal Investigator may have other research staff members who will perform some of the procedures.

You may have questions that this form does not answer. If you do, feel free to contact the Principal Investigator. You may have questions later and you should ask those questions, as you think of them. There is no time limit for asking questions about this research.

You do not have to take part in this research. Take your time and think about the information that is provided. If you want, have a friend or family member go over this form with you before you decide. It is up to you. If you choose to be in the study, then you should sign the form when you are comfortable that you understand the information provided. If you do not want to take part in the study, you should not sign this form. That decision is yours and it is okay to decide not to volunteer.

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**Why is this research being done?**

The purpose of this research is to investigate what African-American college students know about sickle cell disease and sickle cell trait and to determine whether they know their own sickle cell trait status. The decision to take part in this research is yours to make. By doing this research, we hope to learn more about what individuals know about these conditions.

**Why am I being invited to take part in this research?**

If you volunteer to take part in this research, you will be one of about 250 people to do so.

**What other choices do I have if I do not take part in this research?**

You have the choice of not taking part in this research study.

**Where is the research going to take place and how long will it last?**

The research procedures will be conducted online. The total amount of time you will be asked to volunteer for this study is 15 minutes.

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**What will I be asked to do?**

You are being asked to complete a survey consisting of four short questionnaires. You will be asked to answer questions about sickle cell disease and sickle cell trait, answer questions about whether you or any of your family members have sickle cell disease or sickle cell trait, and provide some basic background information about yourself (e.g., name, age, gender). **Since 1987, North Carolina has tested newborns for the presence of sickle cell. If you were born in NC, then by agreeing to participate in this study, you give permission for the researchers to use your name, date of birth, place of birth, and mother's name to access your sickle cell status in the NC State Laboratory of Public Health database, the database that contains all newborn screening test results.**

**What possible harms or discomforts might I experience if I take part in the research?**

There are always risks (the chance of harm) when taking part in research. The risks associated with this research are no more than what you would experience in a normal life. However, some people react to things differently, so it is important for you to tell us as quickly as possible if you any uncomfortable or other negative feelings or experiences while filling out the questionnaires or after completing the questionnaires.

**Are there any reasons you might take me out of the research?**

During the study, if you decide you no longer wish to participate in the study, you may stop participating by not answering any more questions and closing your internet browser.

**What are the possible benefits I may experience from taking part in this research?**

We do not know if you will get any benefits by taking part in this study. This research is designed to might help us learn more about what African-American college students know about sickle cell disease and sickle cell trait and whether individuals know their own sickle cell trait status. There may be no personal benefit from your participation but the information gained by doing this research may help others in the future. If you were born in North Carolina and would like to know your sickle cell trait status, we would be happy supply you with that information. Having this information about your personal medical history could be helpful to you in the future.

**Who will know that I took part in this research and learn personal information about me?**

All answers provided on the questionnaires will be kept confidential.

To do this research, only the organizations/individuals listed below may know that you took part in this study. With your permission, these people may use your private information to do this research:

- Any agency of the federal, state, or local government that regulates human research. This includes the Department of Health and Human Services (DHHS) and the Office for Human Research Protections.
- The University & Medical Center Institutional Review Board (UMCIRB) and its staff, who have responsibility for overseeing your welfare during this research, and other ECU staff who oversee this research.
- Dr. Tamara Warner, the Principal Investigator, and other investigators working on this project.

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**How will my protected health information be used and disclosed?**

The purpose of the information to be gathered for this research study is to better understand what African-American college students know about sickle cell disease, sickle cell trait, and their own sickle cell status. The individuals who will use or disclose your identifiable health information for research purposes include the principal investigator, Dr. Tamara Warner and her co-researchers. Individuals who will receive your identifiable health information for research purposes include only Dr. Tamara Warner and her co-researchers. **The type of information accessed for this research study includes individual sickle cell trait status (positive or not positive), which will be accessed from the NC State Laboratory of Public Health database.** The information will be used and disclosed in such a way as to protect your identity as much as possible; however, confidentiality cannot be absolutely guaranteed. Someone receiving information collected under this Authorization could potentially re-disclose it, and therefore it would no longer be protected under the HIPAA privacy rules (federal rules that govern the use and disclosure of your health information). There is not an expiration date for this Authorization.

You may not participate in this study if you do not agree to Authorize use and disclosure of your health information. You may revoke (withdraw) this Authorization by submitting a request in writing to Dr. Tamara Warner. However, the research team will be able to use any and all of the information collected prior to your request to withdraw your Authorization.

To authorize the use and disclosure of your health information for this study in the way that has been described in this form, please continue to read the rest of the information contained on this form, then follow the instructions at the end of the form. You will be able to print out a copy of this Authorization for your records.

**How will you keep the information you collect about me secure? How long will you keep it?**

The answers you provide to the survey as well as any information obtained from the NC State Laboratory of Public Health will be entered into a database on a secure server which can only be accessed using password protected computers. Any paper records that are generated will be kept in locked filing cabinets in a locked laboratory belonging to Dr. Tamara Warner. This information will be kept for five years after the completion of the study.

**What if I decide I do not want to continue in this research?**

If you decide you that no longer want to be in this research after it has already started, you may stop at any time. You will not be penalized or criticized for stopping.

**Who should I contact if I have questions?**

Dr. Tamara Warner, the Principal Investigator for this project, is available to answer any questions about this research that you may have now or in the future. You can contact her at [warnert@ecu.edu](mailto:warnert@ecu.edu) or (252) 328-6282.

If you have questions about your rights as someone taking part in research, you may call the UMCIRB Office at (252) 744-2914 (Monday through Friday, 8:00 a.m. to 5:00 p.m.). If you would like to report a complaint or concern about this research study, you may call the Director of the UMCIRB Office at (252) 744-1971.

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**Is there anything else I should know?**

You may wish to take note of your surroundings to insure privacy as you take this online survey. When you finish the survey, you should close the browser (e.g., Internet Explorer, Mozilla).

**I have decided I want to take part in this research. What should I do now?**

Read the statements below and if you agree to participate in this research, click the button below to proceed to the survey. This indicates that you have given consent.

- I have read (or had read to me) all of the above information.
- I have had an opportunity to ask questions about things in this research I did not understand and have received satisfactory answers.
- I understand that I can stop taking part in this study at any time.
- By clicking "proceed," I consent to participate in the research study, which includes use and disclosure of my health information. I know I am not giving up any of my rights.
- I may print out a copy of this consent document and Authorization, which is mine to keep.

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## **Appendix H: Debriefing**

## Debriefing

Thank you for completing the research study “Knowledge of Sickle Cell Disease and Trait Status among African-American College Students.” This study was designed to investigate what African-American college students know about sickle cell disease and sickle cell trait and whether individuals are aware of their own sickle cell trait. We were interested in finding out how much you knew about sickle cell disease and sickle cell trait and whether you are aware of your own sickle cell trait status. We also want to find out if people who know their sickle cell trait status have more knowledge about sickle cell disease and sickle cell trait. Finally, we asked questions to determine where your knowledge about sickle cell came from.

If participating in this study has made you feel uncomfortable or caused some distress, please contact the principal investigator, Dr. Tamara Warner by phone at (252) 328-6282 or by email at [warnert@ecu.edu](mailto:warnert@ecu.edu). Dr. Warner is an African-American licensed psychologist who specializes in working with children, adolescents, and young adults with sickle cell disease. She is available to talk to you and answer any questions you might have. She can also refer you to other resources if you need them.

Another on-campus resource that can help if participating in this study has made you feel uncomfortable is the ECU Center for Counseling and Student Development. The Center offers free confidential counseling to all ECU students. The Center is located on the second floor of the Wright Building, Room 316. The telephone number for the Center is 328-6661. For more information about the center, please visit their website at: <http://www.ecu.edu/studentlife/counselingcenter/>

If you would like information about sickle cell disease, sickle cell trait, you can contact the Sickle Cell Disease Association of America (SCDAA), a national organization which provides educational resources for individuals with sickle cell disease and sickle cell trait. The website for the SCDAA is: <http://www.sicklecelldisease.org/>.

If you have any questions regarding your rights as a participant in this study, please call the Chair of the University and Medical Center Institutional Review Board at (252) 744-2914. Questions about the research itself can be answered by Dr. Tamara Warner, who can be contacted at (252) 328-6282 or [warnert@ecu.edu](mailto:warnert@ecu.edu).

