ASSESSING THE FEASIBILTY AND EDUCATIONAL IMPACT OF PROVIDING SICKLE CELL DISEASE EDUCTION IN BARBERSHOPS AND SALONS IN THE AFRICAN AMERICAN COMMUNITY

by

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Sickle cell disease (SCD) is a severe autosomal recessive blood disorder that affects around 1 in 500 African Americans in the United States. Approximately, 1 in 12 African Americans are carriers of sickle cell trait (SCT). The high prevalence of sickle cell trait carriers highlights the importance of having education and trait testing available for individuals, particularly those, who are of childbearing age. Misconceptions and misinformation about sickle cell disease and sickle cell trait can be the reason for an individual to not be motivated to get sickle cell trait testing. This study was created to provide education to the African American community. A pre-post survey research design was used to evaluate 1) knowledge acquisition and retention among study participants who received genetic counseling in the barbershop and beauty salon; and 2) the feasibility of delivering a health education module on sickle cell disease in barbershops and salons in the African American community. As a result of this project, knowledge of sickle cell disease, concern over trait testing, and attitudes toward receiving genetic counseling in a non-clinical setting were evaluated. The data collected suggested that there was significant knowledge gain in 7 of the 9 knowledge questions administered with a p-value <0.05. Concern about trait testing remained low for both the pre and post questionnaires. Attitude about trait testing and genetic counseling remained high for both the pre and post questionnaires as well, resulting in an overall supportive attitude about trait testing and genetic counseling. The mean amount of knowledge gain overall knowledge questions were evaluated

using a paired t-test with significance ($p<0.05$). Overall knowledge gain had a significance of ($p<0.001$). The public health relevance from the results of this study can inform development of health education materials on genetic disorders common in the African American community. Medical and public health professionals can use the insights gained from this study to provide better education and outreach to the community through non-clinical settings in the community.

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1.0 INTRODUCTION

Sickle cell disease is a genetic disorder that more commonly affects those of African descent with an incidence of 1 in 500 African Americans affected with the hematologic disorder in the United States. Since the frequency of disease is higher among this population, carrier screening for this population can provide information to individuals who are at risk. Educating the public about the risks associated with sickle cell trait can help individuals make informed decisions about trait testing and reduce the larger population of African Americans that enter child-bearing age with no knowledge of their risk. This study aims to evaluate feasibility of delivering health education about sickle cell disease in barbershops and beauty salons in the African American community and assess the impact of knowledge acquisition and retention among study participants who receive genetic counseling at the barbershop and beauty salon. This study promotes public health education on sickle cell disease and sickle cell trait to inform the target community.

Using a pre-post questionnaire research design given to the participants of the study at the given barbershop and beauty salon the feasibility of sickle cell disease community outreach and gain and retention of knowledge about sickle cell disease and sickle cell trait were assessed. A total of three questionnaires were administered to participants. The process included a pre

questionnaire, education, and then post and 1 month follow-up questionnaire for statistical analysis of significance.

This study revealed that the educational intervention given at the barbershop and beauty salon is effective at increasing knowledge about sickle cell disease and sickle cell trait ($p<0.001$) and effective at improving the understanding of the role of a genetic counselor to participants ($p<0.001$). Participant's had a high acceptance of encouraging their partner to be tested for sickle cell trait if they were found to be a trait carrier ($p=0.012$) and support for sickle cell disease carrier testing for communities ($p=0.015$) from pre to post questionnaires. Participant's level of concern over trait testing remained low over the courses of the study ($p=0.315$).

The setting of a barbershop and beauty salon can be used for medical outreach for sickle cell disease to increase knowledge about sickle cell disease and sickle cell trait. Education at the barbershop and salon can improve informed decisions making skills and can therefore improve acceptance of sickle cell disease trait testing in communities. Results from this study can help create the development of health education materials on genetic disorders common to the African American community. Future education on sickle cell disease can utilize the insights gained from this study to provide better education and outreach to the community through non-clinical setting like the barbershop or beauty salon.

2.0 BACKGROUND AND SIGNIFICANCE

2.1 GENETIC EDUCATION

Education is one of the most important components of public health outreach in the community. Without education, individuals are unable to make informed decisions and therefore are not autonomous. Even in the area of genetics, education enables individuals to make informed decisions regarding testing or understanding of their genetic make-up. Past community outreach programs involving genetically related conditions have found this to be an important part of a successful outreach project.[1] While many programs have proven to be successful, the first genetic education outreach programs faced much trail. [2; 3] We can use these past experiences to look towards building new projects that are effective for providing education in today's society.

In recent years, there has been a growing call for more public education in genetics.[4; 5] With a better understanding of genetic conditions and their implications on certain populations, the importance of genetic education and expanding genetic knowledge in the community is vital. Studies have been carried out looking at the public's understanding of genetics and genetic literacy. Indications about public illiteracy exemplifies that if individuals have illiteracy in human genetics, it diminishes these individual's opportunity to apply their knowledge to their healthcare.[6] There are a number of reasons why it is important for the public to understand genetics and five of these concepts are outlined by Griffiths: 1) genetics affects one's

worldview, 2) genetics can give insight to crucial issues in society, 3) society depends on genetics, 4) a proportion of human illness has a genetic basis, and 5) genetics provides a classic examples of logical reasoning.[7] The first of these reasons describes how genetics can affect one's worldview and an individual's perception of their biological world around them. Genetics gives individuals insight to not only science, but to how specific traits may be passed to them such as their blood type. Secondly, genetics influences society as seen with discrimination based on sex and ethnic background. In today's society, growing knowledge and understanding of genetics, has made discrimination based on genetics is less common. With the advent of GINA, the Genetic Information Nondiscrimination Act, there is protection for Americans against discrimination based on their genetic information pertaining to health insurance and employment. Thirdly, society depends on genetics for helping individuals understand that they have no control of their genes or how they are passed. Genetics provides the reasoning of why certain individuals may or may not be affecting with a genetic condition or genetic susceptibility. Fourthly, genetics is uncovering that more disorders and human illnesses have an underlying genetic cause. The fifth reason describing why it is important for the public to understand genetics is that it can provide classical examples of logical reasoning for the background basis of how genes are passed downed from generation to generation or how certain people are affected with a genetic condition in a family.

As described above, genetic education provides individuals and society with the autonomy to make informed decisions. Informed decisions are important for any genetic test including carrier screening so that individuals can make the best decision about testing for themselves. Various ethnicities have a higher carrier frequency of certain genetic conditions. These ethnicities are often targeted for outreach. Examples of this can be seen in sickle cell

disease among African Americans, cystic fibrosis among those of European descent, and Tay-Sachs disease among Ashkenazi Jews. Since these conditions are inherited in an autosomal recessive pattern, the increased number of those affected in these populations raises concern about the number of individuals that are carriers in these populations. Carriers are "silent" referring to not exhibiting features of the disease; therefore, family histories are often "silent" as well. When two parents are carriers they have a 1 in 4 (25%) chance of having a child with disease. Being able to target these populations and provide education about risks about carrier status or having a child with one of these conditions can provide some effect on disease prevention from a public health standpoint. It also informs the public on genetic conditions that are more common to their background. Simple education can provide information to individuals that can influence disease outcome.

Genetic conditions such as sickle cell disease, cystic fibrosis, and Tay-Sachs disease are some of the most common conditions where genetic education has had public health significance.[8-12] Educating at risk populations about carrier status, providing them with the information to make informed decisions, and educating them on the benefits and limitations of testing can inform these individuals. The result creates autonomous individuals who can make informed decisions. This educates them to know their risk and have the knowledge to act on it.

A number of education and screening programs have been implemented in the US and around the world. Insights in ways to make an effective outreach project have been studied and written in the literature. Identifying the population and locations of need for the outreach project is key to success.[8; 10; 13; 14] Researching the background and understanding the health beliefs of the target population can make the transition into the community smoother.[15] This can also help with building trust within the community. Empowering participants who receive education can

promote individuals to take an active role in their health by being informed. A study evaluating the health beliefs among African America women regarding genetic testing and counseling for sickle cell disease found a positive correlation between understanding recessive inheritance and perceived susceptibility to having a child with sickle cell disease. Since only half of the respondents understand recessive inheritance, culturally appropriate methods for teaching complex genetic concepts were suggested.[15] Lay health advocates from the target population can provide an additional level of support and education to individuals. Those that receive education are informed and can go out and disperse it to their peers. Discussing how education can be a conversation instead of a lesson can be used to break the barriers that can be perceived with genetic information. An outreach study by Choy et al., outlined specific strategies for developing an effective community outreach program: 1) trust, 2) low key entrée, 3) having a useful approach 4) presentation of appropriate material, 5) utilize visual aids, 6) apply knowledge gained, and 7) having the presence of an authority figure such as a doctor or nurse to legitimize the presentation.[10] Great assets to take along with a provider through outreach are sharing the experience, the knowledge, dedication, humility, consistency, and respect for the target population were all described by Choy et al.[10]

A longitudinal study completed by Barlow-Stewart et al., found high retention of knowledge, low concern, high levels of satisfaction following Tay-Sachs and cystic fibrosis education in high schools.[8] The study emphasized education, and educated participants on inheritance patterns, mutation carrier frequencies, impact of genetic carrier testing, as well as systems for result reporting with the conclusion that the education facilitated informed choice regarding testing and discussion of the implications of its use, and enables knowledge to be retained for the future.[8] Good knowledge and a positive attitude were found to be important

contributors to enabling informed choice about testing.[8; 16] This study proved to successfully evaluate knowledge retention and feasibility of providing education in a school setting. Lessons gained from this project and others can aid in the development of future community outreach projects with genetic education. These topics are just some of the many suggestions that can build a successful outreach project for genetic education whether pertaining to Tay-Sachs or other genetic conditions such as sickle cell disease. Before an outreach project can begin, a significant amount of background research should be carried out in order to address some of the topics discussed in this genetic education session such as disease etiology, disease significance, and disease impact. Described below is a protocol for a community outreach and education on sickle cell disease.

2.2 SICKLE CELL DISEASE

Sickle cell disease is an inherited disorder that affects the red blood cells causing the conformation of their shape to change. It is an autosomal recessive hematologic disease that results from mutations in both copies of the β-globin gene, a subunit of hemoglobin. The pathology of sickle cell disease is complex, varying from moderate to severe anemia, diminished immune function, organ damage, and growth and development delay[17]. Vaso-occulsive "crisis" or pain crisis affects the course of disease in many patients causing pain in the legs and arms, back, chest, or abdomen[17]. Also affected are the cardiovascular, pulmonary, and renal systems[17]. Bone and eye disorders may result as well[17].

Sickle cell disease primarily affects people of African American descent, but it is known to be a global health problem. With the frequency of African Americans affected around 1 in

500 and 1 in 12 for carrying sickle cell trait, sickle cell disease greatly impacts this population.[18] The disease prevalence and the carrier frequency are among the highest seen in genetic conditions. Those that carry sickle cell trait have only one mutation in one of the two copies of the β-globin gene. They are healthy and do not develop sickle cell disease. The important implication that goes along with having sickle cell trait is the risk for having a child with sickle cell disease if both partners have the sickle cell trait. This risk is a 1 in 4 chance of having a child with sickle cell disease, a 1 in 2 chance of having a child with sickle cell trait, and a 1 in 4 chance of having a child without disease or trait. Lack of education and testing for sickle cell trait is one reason that this disease so severely affects this population causing increased morbidity and mortality, both in the United States and globally.

2.2.1 History

Historically, sickle cell disease has been known for generations in Africa under various names.[17] James B. Herrick, a cardiologist at Rush Presbyterian Hospital in Chicago, published the first case report in the United States in 1910 describing the physical characteristics of sickle cell disease as observed in one of his patients.[19] The patient was a 20 year old male who presented to the hospital years earlier with a leg ulcer, and, later pneumonia. On observation of the patients peripheral blood smear, intern Ernest Irons described the anemia and crescent-shaped erythrocytes as sickle shaped red blood cells[17]. Today this is the same term used to refer to the classic description of sickle cell disease. Herrick believed that the abnormal cells may be playing a role in the patient's illness[20].

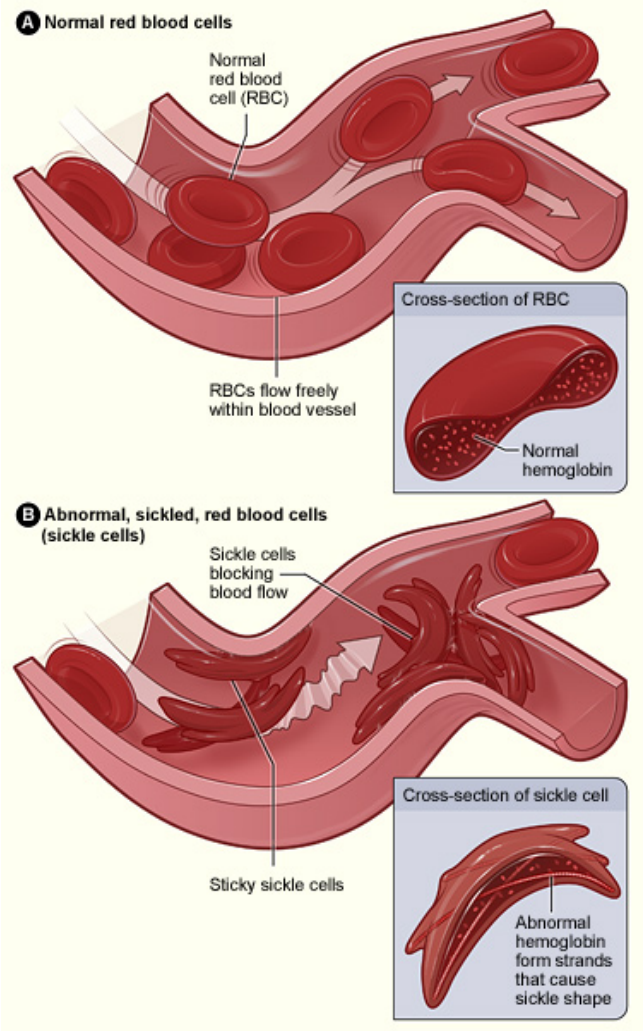


Figure 1: Red Blood Cells (Normal and Sickle Shaped):

A) Depicts a normal red blood cell moving through a blood vessel. The cross-section displays the appearance of normal hemoglobin. B) Shows abnormal sickle red blood cells blocking the blood vessels. The cross-section displays the abnormal hemoglobin that results in the sickle shape of the cells.

(http://www.nhlbi.nih.gov/health/dci/Diseases/Sca/SCA_WhatIs.html 2009.)

The mid-twentieth century led to an over-whelming discovery in sickle cell disease. In 1949, Linus Pauling and his colleagues displayed the electrophoretic abnormality in sickle hemoglobin (HbS) which migrated differently than that normal hemoglobin in an electric field. This led to the realization that sickle cell disease must be related to the hemoglobin molecule[21].

Those with sickle cell disease only had sickle hemoglobin where as those with sickle cell trait had both sickle and normal hemoglobin. Further studies of hemoglobin led to the discovery of other variants of hemoglobin including hemoglobin C, D, and E presenting with milder forms of the disease. Overall, more than 1,300 variants have been identified most showing no clinical significance[22]. Researchers James Neel and E.A. Beet reported the mode of inheritance as recessive.[17] Vernon Ingram, in 1957, attributed the molecular abnormality of sickle cell disease to a substitution of the amino acid valine for glutamic acid at the sixth position of the β-globin chain[23]. The 1970's brought on new technology in the form of recombinant DNA technology and nucleic acid sequencing which provided new insight into sickle cell disease and thalassemia, another inherited hematologic disorder. Nucleic acid sequencing illustrated that the nucleotide change in the DNA for sickle hemoglobin resulted in an A to T change in DNA, a single gene mutation which changed the codon GAG (glu) -Glutamine to GTG (val) - Valine. [17] (See Figure 2) This finding led to the sickle cell disease being the first human disease to be defined by a single nucleotide mutation in the gene[17]. Its discovery developed the foundation for the understanding of protein abnormalities caused by single gene disorders leading to the discovery of many other genetic conditions. This opened the door for sickle cell disease to be the first molecular disease targeted program.

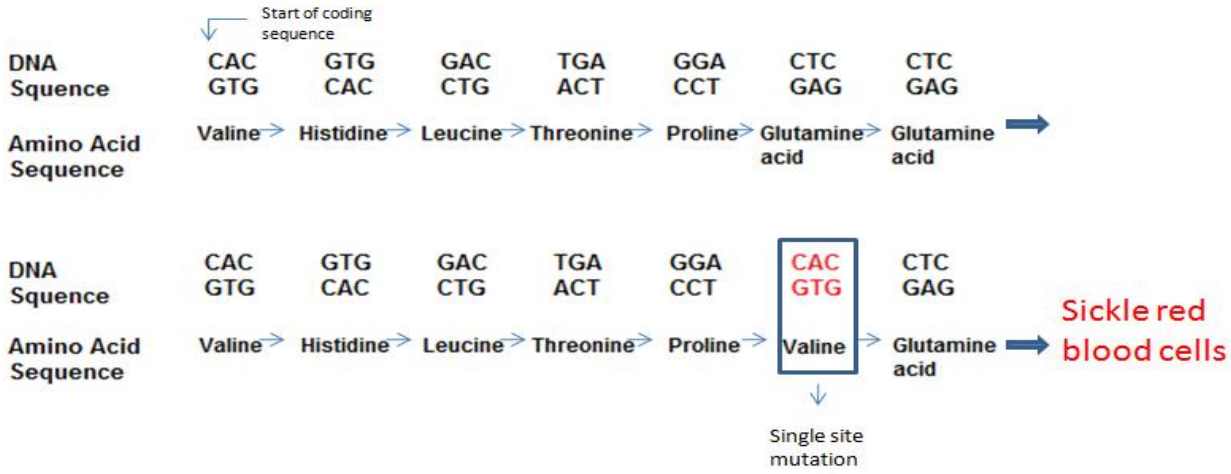


Figure 2: Change in Amino Acid Sequence that Causes Sickle Cell Disease

2.2.2 Molecular Genetics and Pathogenesis

Since the time of Herrick, the understanding of sickle cell disease has come a long way. The knowledge gained from the fore-father of sickle cell disease helped shape the advances that have been made thus far in the understanding of sickle cell disease. Many of the complications associated with sickle cell disease are attributed to the abnormal polymerization of the red blood cell in those with disease. This characteristic has an underlying genetic cause. Mutation in the *HBB* hemoglobin gene, cause the various forms of sickle cell disease which pertains to disease related sickling of the red blood cells including hemoglobin SS, hemoglobin SC, hemoglobin SD, and two forms of beta-thalassemia HbS β^{+} and HbS^{0}.[24] Sickle hemoglobin is less soluble than normal hemoglobin upon deoxygenation. The sickle hemoglobin forms fibers within the red blood cells, leading to morphological changes resulting in occlusion of small blood vessels with acute pain and chronic organ damage.

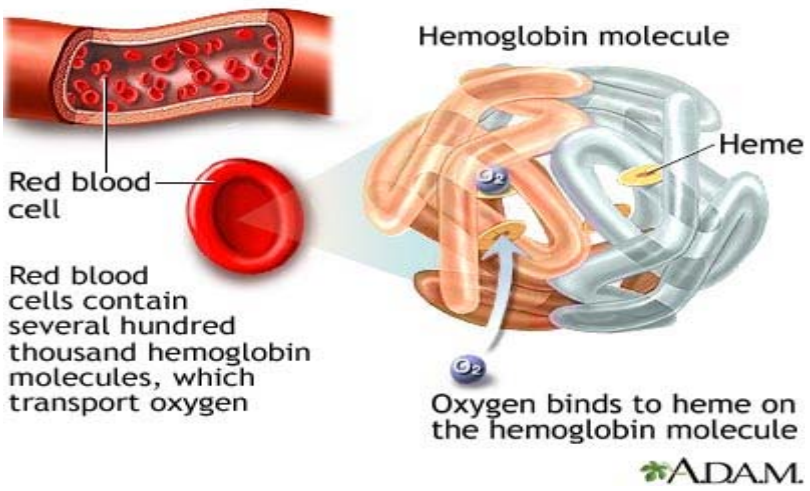


Figure 3: Hemoglobin Molecule

(http://www.nlm.nih.gov/medlineplus/ency/imagepages/19510.htm)

The β-like gene cluster located on chromosome 11 encodes the following five functional genes ε, $^{G}\gamma$, $^{A}\gamma$, δ, and β-globin expressed during development.[26] The α-like globins gene ζ_1, ζ_2, α_1, and α^2 are found on chromosome 16. Hemoglobin S as stated previously is caused by the substitution of valine for glutamic acid on the sixth position of the β-globin gene resulting in homozygous HbSS disease causing 65% of sickle cell disease. The compound heterozygote state HbS and HbC represents 25% of sickle cell disease, 9% caused by SCD-Sβ thalassemia and the remaining one percent consisting of other variants (SCD-SE, SCD-SD^{Punjab} , SCD-SO^{Arab} **,** and SCD-S^{Lepore}).[27] Thalassemias related to α-like gene deletions called α-thalassemia cause another type of hemoglobin disorder. Deletions in the α-globin gene include one-gene deletion silent carrier state (/-α) and a two-gene deletion (-α/- α) producing α-thalassemia trait. Alpha-thalassemia most commonly affects those of Asian descent.

The pathophysiology of sickle cell disease is very complex given the number of diverse clinical phenotypes of the disorder. From patient to patient the presentation of sickle cell disease varies. Some patients may present with stroke, acute chest syndrome, and vaso-occulsion while others will just have a small number of pain episodes that don't require hospitalization. The

pleiotropy of the disease leads to the care of patients to be individualized depending on presentation of features. The most common features of sickle cell disease were indentified through a study called the Cooperative Study of Sickle Cell Disease (CSSCD) examining the natural history of the condition.[28-30] The most common features identified were painful episodes, acute anemic episodes, acute chest syndrome, complications related to the central nervous system including stroke and cerebral ischemia, priapism, growth and development delays, and other complications.[28]

2.2.3 Inheritance and Incidence

Sickle cell disease is a genetic disorder with an autosomal recessive mode of inheritance and follows the rules of Mendelian Genetics. Autosomal refers to the 22 autosomal chromosomes which are the same in both males and females. Recessive means that both copies of the β-globin gene need to have a mutation present in order for an individual to be affected with sickle cell disease. Therefore, if each parent is a carrier of sickle cell trait (AS), then with each pregnancy there is a 25% chance that their child would have sickle cell disease (SS), a 50% chance their child would carry sickle cell trait (AS), and a 25% chance their child would have two working copies of the gene (AA). All children of an affected individual with sickle cell disease will inherit the mutation of their parent and be carriers.

In the United States, sickle cell disease is most common among African Americans. Every year, approximately 1 in 500 African American infants are born with sickle cell disease from inheriting the β^s allele from both parents. The carrier frequency of sickle cell trait in African Americans is approximately 8%.[25] About 8% of African Americans in Western PA are carriers of sickle cell trait. Since admixture is occurring more commonly sickle cell disease

alleles are becoming somewhat more distributed among populations. The prevalence rate of all forms of sickle cell disease in individuals of African American descent is around 85%.[25] Secondly, Native Americans are the next most commonly affected at a 10.6% prevalence rate.[25] Asians, Hispanics, and Caucasians make up the remaining groups with a prevalence rate of 4.4%.[25]

2.2.4 Clinical Manifestations of Sickle Cell Disease

The basis of sickle cell disease is a single beta-globin mutation coding for the sickle beta-hemoglobin. When a person carriers two of these mutations, the hemoglobin can take on an abnormal shape, distorting the shape of the red blood cell. These cells change from a normal round, donut shape to the elongated "C" shape of a sickle red blood cell. Sickle cells have a shorter-than-normal lifespan, which leads to anemia (low red blood cell count). A normal red blood cell lives for 120 days, whereas a sickle cell lives for only 10-20 days.

Most children with sickle cell disease have some degree of anemia and might develop one or more of the following conditions and symptoms as part of the disorder. The most common symptom identified is pain. It is defined by the CSSCD as a painful episode lasting at least two hours, unexplained by any medical condition, and requiring medical attention.[28] A pain "crisis" episode may be triggered by febrile illness, dehydration, stress, and extreme environmental temperature changes.[31] In some cases there may be no event that can be identified as the cause of it. In a study by Gill et al., painful episodes mainly occurred in sickle cell disease-SS and sickle cell disease-Sβ^0 thalassemia at a rate of 80 and 100 episodes per 100 pt-years.[28] Children ages 0-9 years have lower pain rates than those older than 10 years .[32]

Secondary to pain is acute splenic sequestration, parvovirus B19 infections, sepsis or folate deficiency which results in acute anemia.[31] Acute anemia is defined as a precipitous drop in the red blood cell count and a reduction of hemoglobin or hematocrit of 30% below steady-state values unrelated to other known SCD-related causes.[33]

Acute chest syndrome ACS is another commonly occurring symptom of sickle cell disease. It is defined by the CSSCD as an episode causing a new infiltrate on chest x-ray or a demonstrated pulmonary perfusion defect on radioisotope scan. There are many factors that can be attributed to ACS including genotype (sickle cell disease-SS and sickle cell disease-$S\beta^0$ thalassemia), young age, higher Hb and leukocyte counts, and low HbF.[31] A study researching the high frequency of children with ACS found that these children commonly present with signs of infection, whereas adults get ACS commonly after hospitalization from another complication of sickle cell disease.[34] ACS can also be caused by rib infarcts or injury to lung tissue. [35]

Complications found in the central nervous system are seen in those with sickle cell disease. Increased mortality in children with sickle cell disease causes complications of stroke or cerebral vascular accident.[31] Strokes are described as acute neurologic events due to vaso-occulsion or hemorrhage associated with cerebral ischemia and neurologic signs and symptoms.[36] They can result from moyamoya or rupture of the arteriovanous malformations and are most commonly seen in patients with sickle cell disease-SS and sickle cell disease-$S\beta^0$ thalassemia.[31] Around 14% of patients who have a stroke have a recurrence of stroke between 3 to 22 months of time.[31] There are a number of risk factors for stroke including a history of transient ischemic attack, ACS within two weeks of acute stroke, a high frequency of yearly ACS, high systolic blood pressure, low stead-state Hb, and increased WBC counts.[37] Through

trans-cranial Doppler (TCD) studies, patients with sickle cell disease can be monitored in order the identify those at an increased risk for stroke.

Those with sickle cell disease have been identified with "silent" central nervous system (CNS) infarcts. They are described as ischemic changes on brain magnetic resonance imaging (MRI) scan happening on patients who have not had a history of stroke.[38] These children have been observed to have higher rates of abnormal neuropsychological studies.[38] Higher rates of "silent" infarcts have been associated with a history of pain "crisis" episodes, a history of seizures, WBC counts over 11.8 x 10^9 per liter, and Sengal β-cluster haplotype.[39]

Other complications that are more common to those with sickle cell disease in priapism, avascular necrosis, and growth and development delay. Priapism is commonly seen in males with sickle cell disease. It is a painful penile erection that lasts longer than one hour requiring medical attention. High lactate dehydrogenase levels and reticulocyte and platelet levels were all associated with priapism.[40] Treatment with hydroxyurea has shown to have positive benefits to males. Avascular necrosis (AVN) of the hips affects some with sickle cell disease. They can sometimes lead to chronic pain "crisis" episodes. Delayed growth and development have affected children with sickle cell disease. This delay also includes pubertal development. It is more identifiable in those with sickle cell disease-SS than sickle cell disease- SC.[31] Menarche is delayed by two or three years with a mean age around 15-16 years.[41] Children receiving transfusions may have improved growth in height and weight compared to those who do not received transfusions.[31] The complications of sickle cell disease affect all areas of the body. They are often hard to predict and treatment for sickle cell disease is most often based on manifestations of symptoms.

2.2.5 Diagnosis

Sickle cell disease is most commonly diagnosed in infancy in the United States. All newborns in the US are screened for hemoglobinopathies through newborn screening, and approximately, 2,000 infants with SCD are identified annually.[42] Screening is predominantly with the use of isoelectric focusing (IEF) which identifies infants with other hemoglobinopathies and hemoglobinopathy carriers. The diagnosis is established by demonstrating the presence of some form of disease either Hb SS or other abnormal beta globin chain variant. In the event that a screen received is positive, the standard of care for a baby with disease is confirmatory testing of a second blood sample should be completed by 2 months of age so that parental education, prophylactic penicillin, and comprehensive care can be initiated.[43; 44] Confirmatory testing is done through hemoglobin electrophoresis.

2.2.6 Management of Sickle Cell Disease

Management of sickle cell disease is based on clinical manifestations of the disease for each individual. Initially, after a diagnosis is made parental education, prophylactic penicillin, and comprehensive care begin. Education should include information about the specific type of sickle cell disease that affects the child. Education is not routinely accomplished in one session. Future sessions are planned to provide information appropriate for the child's age and diagnosis. Evaluations to monitor the child's physical health routinely are necessary for tracking the course of each individual child's disease. This includes routine physical examinations, laboratory tests, and brain and lung imaging.

Major complications of sickle cell disease are handled on a case by case basis depending on the clinical manifestations of the individual. Educating parents about knowing the signs of a problem are critical in successful management of a child with sickle cell disease. This also goes for adult sickle cell disease patients. The most commonly treated problems are fever, pain episodes, acute chest syndrome, and stroke.

Fever is one of the most common signs of illness caused by infections in children. Pneumococcal infection and penicillin prophylaxis have reduced the risk of mortality of sickle cell disease patients from infection, and they are an integral part of medical management of patients with sickle cell disease. All children who have sickle cell disease who have fever (101^0F) and other signs of infection should be evaluated promptly.[45] If an infection is present then broad-spectrum antibiotics should be given.[45] Acute splenic sequestration and erythroid aplasia "aplastic crisis" occur commonly with fever.

Pain episodes are one of the earliest complications seen in sickle cell disease patients in the form of dactylitis "hand-foot syndrome". Vaso-occlusive pain may involve the limbs, abdominal viscera, ribs, sternum, vertebrae, and sometimes skull bones.[45] Depending on the severity of pain various medications can be used for pain relief. Mild to moderate pain are managed with acetaminophen and non-steroidal anti-inflammatory drugs, such as ibuprofen, and mild opioids, such as codeine.[45] Stronger non-steroidal anti-inflammatory drugs and opioids are reserved for those with severe pain.[45]

Treatment of acute chest syndrome includes an assessment of blood oxygenation, simple transfusions, intravenous broad-spectrum antibiotics, and pain control. Administration of oxygen to moderately hypoxemic patients can be of benefit to patients experiencing acute chest syndrome. Transfusions decrease the proportions of sickle red blood cells and increase the

oxygen affinity of the blood. Antibiotics given to febrile or severely affected acute chest syndrome patients because of the challenge it is to exclude bacterial pneumonia or superinfection of a lung infarct.

Stroke management is carried out with the use of Transcranial Doppler (TCD) imaging and blood transfusions. In rare instances, sickle cell disease has been cured through bone marrow or stem cell transplant.[46] It has been successful with minimal recurrence and mortality. Approximately, 85% of children survive free of the underlying disease after human leukocyte antigen (HLA)-identical sibling donor hematopoietic cell transplantation (HCT).[47]

2.3 SCREENING

2.3.1 Newborn Screening for Sickle Cell Disease

Newborn screening has two main purposes according to the American College of Medical Genetics 1) to screen for genetic disease or genetic predisposition to prevent the effects of the disease and 2) to retrieve, diagnosis, and provide intervention before irreversible damage occurs.[42] In order to do this, there should be a foundation for a successful screening program. The elements of this foundation should include that there be an identifiable population at risk, that the screening test should be technically simple, inexpensive, and suitable for mass processing, that there must exist a reasonable form of treatment or prevention for the disease being screened, and that the test reliability should be high.[48] In the state of Pennsylvania currently there are six genetic conditions screened on the panel: phenylketonuria, congenital

hypothyroidism, hemoglobinopathies, galactosemia, maple syrup urine disease, and congenital adrenal hyperplasia.

Hemoglobinopathies have been included in newborn screening in the United States since the 1970s.[49] Screening for sickle cell disease in newborns currently is mandated in all 50 states and the District of Columbia.[50] It was adopted in 1996 when the American Academy of Pediatrics recommended that every state implement targeted newborn screening for hemoglobinopathies.[51] Most states either use thin-layer isoelectric focusing (IEF) or high-performance liquid chromatography (HPLC) for screening procedures.[50] Both have a high sensitivity and specificity. The primary purpose of newborn screening for sickle cell disease is to identify newborns with disease, and initiate penicillin prophylaxis for the purpose of prevention of mortality from pneumococcal sepsis and splenic sequestration.[42] Penicillin prophylaxis significantly reduces the incidence of pneumococcal sepsis in infancy a common cause of mortality in infants. It is standard of care to beginning penicillin prophylaxis at 2 months of age and immunizations at recommended intervals.[50] In the course of screening for sickle cell disease, heterozygote carriers of sickle cell trait are identified as well. This information offers the opportunity to counseling families that could be at risk for having a child with sickle cell disease in a future pregnancy.[15]

Since the beginning of newborn screening and something that is true today are problems with follow-up for those that screen positive for sickle cell trait. In 1987, the NIH created a consensus statement on newborn screening for sickle cell disease and other hemoglobinopathies. The statement addressed the efficacy of newborn screening for sickle cell disease, techniques, factors to be considered, optimal follow-up and management, and future research decisions.[52] The committee found that effective intervention of children with sickle cell disease provides a

major impetus for neonatal screening since reliable, simple, and inexpensive techniques for mass screening are available. The apparent benefits from screening permit state law mandate of these services while permitting parental refusal. In order to be effective, screening must be part of a comprehensive program for the care of sickle cell patients and their families. This should include optimal medical care, psychosocial support and genetic counseling. Further research should focus on improving and evaluating technology for screening; defining the impact of screening on the physical, social, cognitive, and emotional development on the child and on family members; assessing other methods of management of infection; and providing optimal education of individuals and families at risk.[52] Even twenty years after this time, issues still arise regarding the newborn screening guidelines.

2.3.2 Knowledge and Perceptions about Sickle Cell Disease and Sickle Cell Trait

It is particularly important for individuals with sickle cell trait to be aware of their risk to have a child with disease in the future. Given the high prevalence of sickle cell trait in people of African descent, understanding the knowledge and perceptions of those individuals that are at increased risk for sickle cell disease and sickle cell trait is beneficial. There has been limited research on the subject and past research has been influenced by misinformation and misconceptions on the topic.[2; 3]

In a study of 147 African American patients ages 18-50 seen in an emergency department, found that 73% knew that sickle cell disease was a genetic disease.[12] Of this sample, only 31% actually knew their own trait status.[12] Another study showed that 23% of a sample of patients with sickle cell disease incorrectly thought that sickle cell trait can change

into sickle cell disease.[53] These data suggests that there is a lack of awareness about sickle cell disease even among those at greatest risk.

Public education on sickle cell disease has been limited and limited studies have been carried out looking at the effects of public education on the community.[8; 10; 12; 54] In the 1970s, coercive reproductive politics and insurance and employment discrimination were associated with sickle cell trait screening. The African American community associated sickle cell disease testing and follow-up with distrust. Many referred to it as "the black disease." With this past history, implementing new strategies for sickle cell disease and sickle cell trait outreach should be centered on beneficence to the individual and be particularly sensitive to those serve the target population and be culturally appropriate.

In a study by Treadwell et al., qualitative and quantitative strategies were used to delineate general level of awareness and understanding of sickle cell disease and sickle cell trait in the community to help better understand and address the knowledge and perceptions of sickle cell disease and sickle cell trait in the community.[54] The study found common themes across focus groups including limited general awareness of sickle cell disease and sickle cell trait, to much emphasis on the nature of the disease instead of the future implications of sickle cell trait, and the need for public health education about sickle cell disease and sickle cell trait involving the media.[54] The study found that there was good general knowledge of sickle cell disease, but this did not impact the percent of people that actually knew their trait status.[54] They also found that if respondents had received information from friends or acquaintances they were three times more likely to know their trait status. In conclusion, knowledge of sickle cell disease was entering the mainstream; however, this knowledge was not increasing the number of people who know their sickle cell trait status. Treadwell also discovered that despite past controversy with

sickle cell trait, public health education can be a model for the empowerment of communities in making informed decisions and aiding in family planning.

A study evaluating the knowledge and health beliefs of sickle cell disease and sickle cell trait on the acceptance of genetic screening for sickle cell trait found that education of sickle cell disease in a prenatal setting can influence knowledge of sickle cell disease and the acceptance of genetic testing for sickle cell trait. According to the study that utilized the health belief model to evaluate health beliefs, surveyors felt that they had a low belief that individually they were at risk for being carriers of sickle cell trait.[55] The study concluded that simple education can be a success, but in order to have maximum success educational materials need to be sensitive to the beliefs of the population that are leading to a lack of motivation for testing particularly the beliefs that they are not at risk for sickle cell disease or sickle cell trait.[55]

2.3.3 Barbershop Outreach

One way to impact the knowledge and perceptions of the public about sickle cell disease and sickle cell trait is to go into their community in a place that is comfortable to them.[56] A number of projects have tried this approach at the church and the barbershop.[57; 58] Barbershop outreach has been carried out in only a limited number of studies.[56-59] Due to the lack of individuals contact with the healthcare system, there is a lack of dissemination of education to those that are at increased risk for certain conditions such as sickle cell disease or sickle cell trait. The church has been one setting that has been somewhat effective for public health outreach for African Americans. However, in looking back at who was reached, regular church attendance is much less common among African American men than women. A way to better target men was attempted at the barbershop.[58]

The barbershop is a cultural institution that attracts a large number of individuals who have a tendency to come to get their haircut on a regular basis for both men and women. It's a place where conversations are open and those present are free to talk and express how they feel about various topics and discuss various topics. Barbershops have been used in previous studies for blood pressure screening and prostate screening.[56-59] This study was based off of the location of the barbershop and the group dynamic that goes on in it so that sickle cell disease and sickle cell trait education could be carried out a novel community setting to accomplish public health education.

3.0 SPECIFIC AIMS

3.1 SPECIFIC AIM 1

Specific Aim: To examine the feasibility of genetic education, genetic counseling, and genetic testing in the community setting of a barber shop and a beauty salon regarding sickle cell disease (SCD) and sickle cell trait (SCT).

Hypothesis: The community setting of barber shops and beauty salons is a feasible location for genetic education, genetic counseling, and genetic testing of SCD and SCT. The high traffic, client based social setting of neighborhood barber shops and beauty salons create a location where education, counseling, and testing can occur.

Plan: Anonymously paired questionnaires including both close and open ended questions pertaining to SCD and SCT education, genetic counseling, and genetic testing in the community will be given in order to examine if genetic education, genetic counseling, and genetic testing for SCD and SCT is feasible in the community based on participants' responses.

3.2 SPECIFIC AIM 2

Specific Aim: To evaluate the impact of genetic education, genetic counseling, and genetic testing in barbershops and beauty salons in the African American community by testing the acquisition and retention of knowledge of sickle cell disease and sickle cell trait and by evaluating study participants' responses in the study questionnaires.

Hypothesis: Genetic education, genetic counseling, and genetic testing in the setting of a barber shop or beauty salon can be an effective way to impact knowledge of SCD and SCT in the community.

Plan: Anonymously paired surveys including statements on sickle cell disease will be used to find if genetic education, genetic counseling, and genetic testing in the setting of a barber shop or beauty salon can impact the acquisition and retention of knowledge of those that receive genetic counseling. The pre-questionnaire will be given before sickle cell disease education then the post-questionnaire will be given immediately following education and one month following education to evaluate the impact that education can have in this community setting to these participants.

4.0 MATERIALS AND METHODS

The study design consisted of a community-based educational strategy to raise awareness of sickle cell disease and sickle cell trait. It was approved by the University of Pittsburgh's Institutional Review Board (IRB) on October 6, 2008 (Replications of the IRB approval letter for protocol # PRO08020016 can be found in Appendix A). Previous to approval, a partnership between with the Sickle Cell Program at Children's Hospital of UMPC and the University of Pittsburgh Center for Minority Health (CMH) were made in order to facilitate this study. A presentation of the study and its purpose were given to the Community Research Advisory Board (CRAB), the advisory board to the CMH, in March of 2008 prior to the beginning the project for consultation for research planning and building cultural confidence.

4.1 PARTNERS

4.1.1 Children's Hospital of Pittsburgh of UPMC

Children's Hospital of Pittsburgh of UPMC and the Sickle Cell Program have been devoted to caring for those with sickle cell disease. Since 1978, Children's has been identifying children with sickle cell disease and provided children and parents with counseling, care, and further genetic testing. The goal of the Children's Sickle Cell Program is to identify children with sickle

cell disease as early as possible and manage their condition. Another aspect of Children's Sickle Cell Program is educational outreach and community awareness. Funding through a HRSA grant entitled "Western Pennsylvania Sickle Cell Network: An Integrated System of Care for the Enhancement of Newborn Screening Follow-Up" (grant number: 6H46MC 00255-01-01) enabled funding for trait testing for study participants who interested trait testing. This was offered separate from the IRB study itself with no data collection of testing results used for research. The Sickle Cell Program collaborated closely with the Center for Minority Health because of the experience and trust established with local barbershops and salons in Pittsburgh, PA.

4.1.2 Center for Minority Health

The Center for Minority Health established at School of Public Health at the University of Pittsburgh in 1994 has a history of commitment to translating evidence-based research into community-based interventions and innovative outreach projects.[59; 60] The center practices addressing health issues among ethnic minorities and other vulnerable and underserved populations. According to the center's vision statement, "The Center for Minority health envisions a society which values and contributes to health, social equity (fairness) and equality (sameness) for all people." The infrastructure contributes to a number of outreach projects serving minorities. The Health Advocates in REACH (HAIR) ™ is a project that provides health related services in barbershops and beauty salons to people in the community.[59] It is aimed at reaching African Americans who continue to suffer from health disparities.[61]

Physicians, nurses, and other health professions in the project entitled "Take a Health Professional to the *People* Day" go into 10 local barbershops and beauty salons in Pittsburgh in

order to provide health screening and education.[60] The idea behind providing healthcare at the barbershop or salon steams from research analyzing the idea that trusted community members such as barbers and beauticians are good vehicles for disseminating information and promoting health messages related to disease prevention.[59] The project has been successfully carried out annually since September 2002. Based on this outreach model, the sickle cell disease educational outreach project in barbershops and beauty salons was developed. .

4.2 PARTICIPANTS

Participants in the study met human subjects' criteria as defined in the IRB protocol of being either male or female and 18 years of age and older. No racial or ethnic subgroups were excluded from participation in the study. All individuals recruited into the study were able to read and comprehend English. The participants were recruited from one barbershop and one beauty salon located in Pittsburgh, PA. Advertisements were not used at the shop for this particular study, but they were used to advertise the sickle cell trait testing days performed in conjunction with the study (See Appendix D). Each participant was approached by the principle investigator (PI) for recruitment. Recruitment was random and based on the criteria as outlined through the IRB protocol. Recruitment, consent, education, and surveys were given by sickle cell disease community educator (principle investigator). After recruitment, each participant was consented for the study, given the pre-questionnaire, education, post-questionnaire, and one month follow-up questionnaire if available. An explanation of the process can be found in Appendix C.

4.3 METHODS OF ASSESSING FEASIBILITY

4.3.1 Specific Aim 1

Specific Aim 1 was designed to examine the feasibility of genetic education, genetic counseling, and genetic testing in the community setting of a barber shop and a beauty salon regarding sickle cell disease and sickle cell trait. Feasibility was assessed based on questionnaires including pre, post, and one month follow-up questionnaires. The questionnaires examined participants' knowledge of sickle cell disease and sickle cell trait, concern over carrier testing for sickle cell trait, and attitudes toward sickle cell disease and genetic screening in the community as well as genetic counseling. The questions used in questionnaires had previously been piloted to evaluate genetic education in a previously published genetic screening program for Tay-Sachs disease and cystic fibrosis by Barlow-Steward et al. and a study examining the knowledge and health beliefs of sickle cell disease and sickle cell trait on the acceptance of genetic screening for sickle cell trait.[8; 15] Based on the participants responses to the knowledge, concern, and attitude statements as well as open ended questions found on the questionnaires feasibility of genetic education, genetic counseling, and genetic testing in the community were assessed (See Appendix B for questionnaires).

Along with the ability of administering the questionnaires and receive responses, the feasibility was evaluated on the continued cooperation of the two participating locations Ms. Ida's Barbershop in Wilkinsburg. PA and A Second Glance in East Liberty, PA. The two locations were selected to provide the range of target population, culture, demographics, and setting for sickle cell disease and sickle cell trait education and pre-post test questionnaire administration. The ability to retain participants through the process of pre-questionnaire,

education, post-questionnaire, and one month follow-up was also a method of assessing feasibility.

4.4 METHODS OF EVALUATING IMPACT

4.4.1 Specific Aim 2

The second part of the study was aimed at evaluating impact of genetic education, genetic counseling, and genetic testing in barbershops and beauty salons in the African American community by testing the acquisition and retention of knowledge of sickle cell disease and sickle cell trait and by evaluating study participants responses to the study questionnaires. Impact was assessed based on questionnaire responses.

In order to evaluate the impact of education, educational material relevant to the knowledge questions was developed. Materials include educational slides and take-home brochures. (See Appendix E) The educational slides were designed based on questions in the surveys pertaining to the knowledge of sickle cell disease. With the use of this design of study questions, Barlow et al., found that there was a high retention of knowledge, low concern, and high levels of satisfaction.[8] A pre-questionnaire was given to each participant to evaluate their knowledge before education. A brief educational session was presented by a sickle cell disease community educator reviewing the natural history of the disease, etiology, and the importance of

understanding sickle cell trait. An outline of the questionnaires including visual aid slides and brochures given to the participants can be found in Appendix B and D.

Immediately after the educational session an identical post-questionnaire was administered to assess the change in knowledge as well as to assess any change in concern or attitude towards trait testing or genetic counseling. The post-questionnaire also included open ended questions pertaining to study participants' perception of receiving health services at a location like a barbershop or beauty salon, if genetic counseling was helpful experience, if they learned something, if they felt like they could talk to other about what they have learned, and if they would recommend a program like this for others. One month after the educational session, participants were given an identical questionnaire in order to assess the retention of knowledge of those that received education and any changing levels of concern or attitude.

Responses of knowledge statements were converted to dichotomous variable where 1 represented correct answers and 0 represented incorrect and uncertain answers. A total of nine knowledge questions were evaluated for impact regarding acquisition and retention of knowledge. The responses to each statement that were converted into dichotomous variables were analyzed using the statistical Fisher's Exact Test to test the association between pre, post, and one month follow-up questionnaires for each statement between each test. In order to evaluate the impact of the overall knowledge questions, concern, and attitude questions. Each category was analyzed using a paired t-test to look for overall knowledge gain, concern, and attitude.

5.0 QUALITATIVE ANALYSIS

Qualitative analysis was carried out in order to analyze the open-ended questions in the post and one month follow-up questionnaires. A total of 6 open-ended questions were used for analysis. These questions explored participates feeling on receiving health services at their local barbershop/beauty salon, their thoughts on genetic counseling, if they felt like they could talk to others about what they learned about sickle cell disease and sickle cell trait, and if they would recommend a program like this for others. These open-ended questions allowed participants the opportunity to voice their feelings and experiences from being in the study. This allowed for new unexpected and unexplored areas of information to be analyzed. Participants could be free from the standardized answer choices given for the quantitative analysis portion of the study and be candid with their responses.[62] As described by Patton, quantitative data is succinct, systematic, standardized, and easily aggregated for analysis.[63] Qualitative findings on the other hand are longer, more detailed, and variable in context.[62; 63] This can make analysis more complex and in depth because responses are neither systematic nor standardized.[63] Qualitative analysis allows the research to capture the view points of participants without predetermining these points through prior selection of these categories.[62]

6.0 DATA ANALYSIS

6.1 FEASIBILITY

Data collection and retention of participants assessed the feasibility of sickle cell disease and sickle cell trait genetic education, genetic counseling, and genetic testing in the community setting of a barbershop and a beauty salon. The IRB protocol allowed for 50 participants to be enrolled in the study. Data was collected on the 50 consented participants. Feasibility of participant retention was based on the percent of participants that remained for the duration of the study. Based on other studies assessing the feasibility of genetic education, their range of retention of participants were between 42.7-88.2%.[8; 9; 64] It was deduced that if this study fell within this range that its participants' retention rate would be considered adequate, and this would make up part of the result of how this project would be considered feasible. Another aspect of the feasibility was through the continued participation of the barbershop and beauty salon which were the two locations of educations.

Feasibility was also assessed on the responses of the study participants. Based on their attitudes and concerns over education, trait testing, and genetic counseling continued feasibility of the study was analyzed. The study participants' responses provided evidence of whether this study was feasible. Completion of questionnaires and responses to open ended questions could conclude this as well.

6.2 IMPACT

The impact of genetic education, genetic counseling, and genetic testing in barbershops and beauty salons in the African American community was evaluated by testing the acquisition and retention of knowledge of sickle cell disease and sickle cell trait and by evaluating study participants responses based on questionnaires. The data collected analyzed the impact of knowledge, concern, and attitude from the questionnaires. Question items from main categories of information (knowledge, concern, and attitude) were analyzed individually and as an overall sum within each category.

For each individual for each of the nine knowledge questions, correct responses were assigned a "1" whereas incorrect "uncertain" responses were assigned "0". Using Fisher's exact tests, the proportion of correct responses were then compared between pre and post-education session, as well as pre and 1-month afterwards. We specifically wished to test whether there was a significant gain of knowledge that is the proportion of correct answers would be higher for the post and one-month follow-up questionnaire. The null hypothesis used to test the significance of knowledge gained was that there was no knowledge gain between the tests. Significance was determined based on a p-value of <0.05. Finally, we calculated a sum of responses across all nine knowledge questions used a paired t-test ($p<0.05$) to determine whether overall knowledge increased post and one-month after the education session.

Concern and attitude were also evaluated for each item individually to determine if there were any differences in concern or attitude before and after education, and then also evaluated them as a whole within each category. Similar to the methods described above, Fisher's Exact Test was used to test null hypothesizes that there was no change in concern or attitude between the questionnaires. First the pre and post were compared, then pre and one month follow-up, and

finally post and one month follow-up. Based on a p-value of <0.05 the responses to the questionnaires were evaluated for significance. Overall significance of concern and attitude responses were grouped together and evaluated using a paired t-test with significance $p<0.05$. Participants' responses to open ended questions were also used to qualitatively analyze impact of education, concern, and attitude.

7.0 RESULTS

7.1 SURVEY RESULTS

7.1.1 Demographics

A total of 50 individuals were consented for the study. Of those individuals 47 completely accomplished the initial pre and post questionnaires and received education. Three of those participants were lost to follow-up. The reasons behind these losses pertained to timing and having to leave the barbershop/salon before the process could be completed. Of the 47 participants that completed the process, 49% were males and 51% were females. The average age range for participants were 31-35 years of age. Participants' marital status was predominantly single (73%). The majority of the individuals had children (63%) and some level of college (29%). When asked about health insurance 85% of participants had some form of health insurance and 15% did not. (See Figures 3-7)

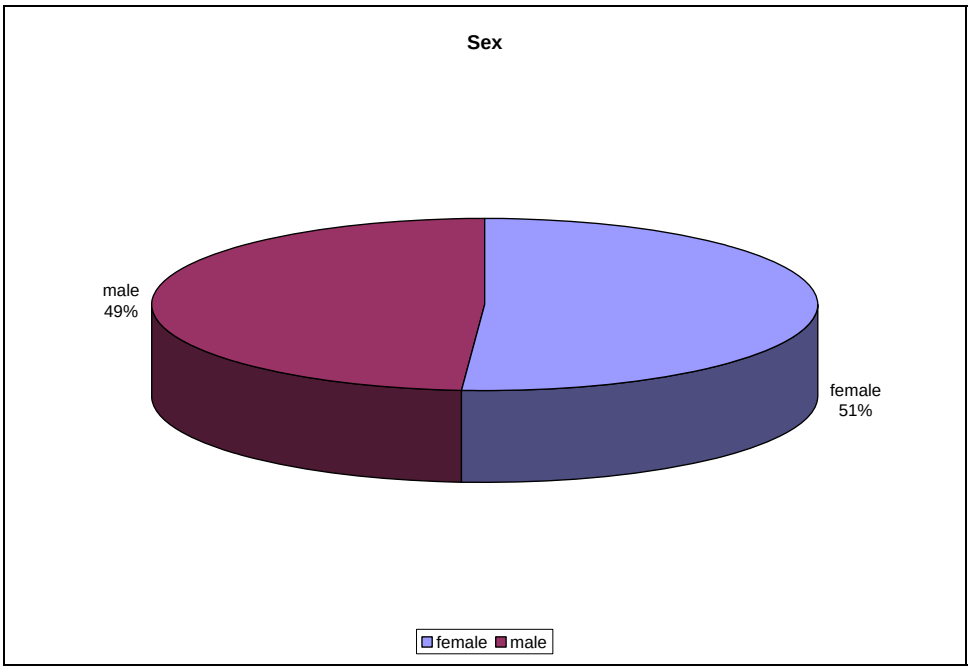


Figure 4: Percentage of Males vs. Females

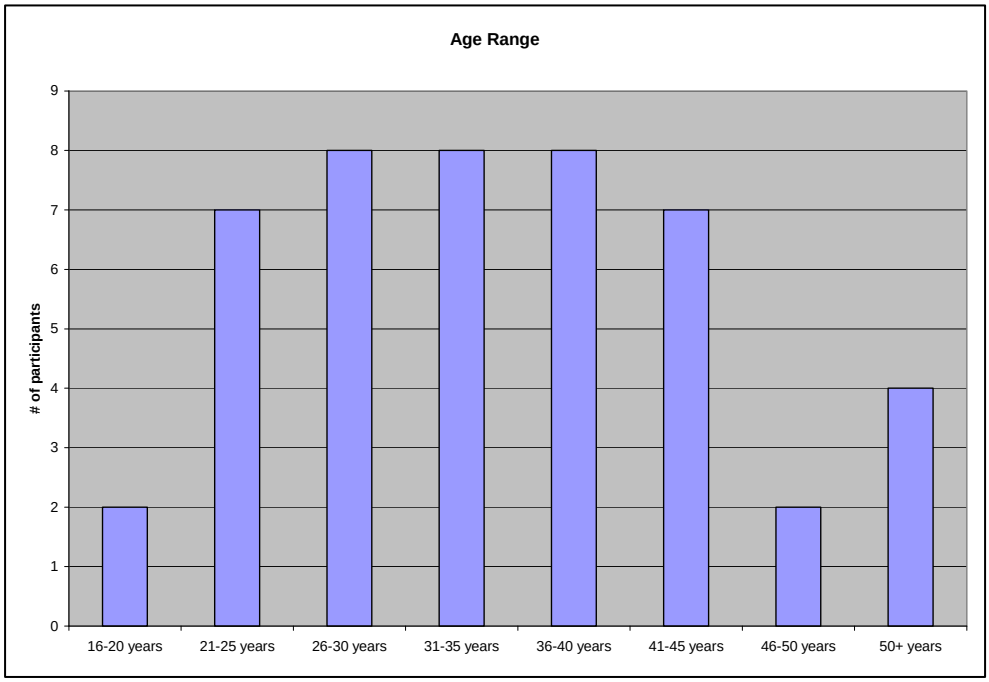


Figure 5: Age Distribution of Survey Participants

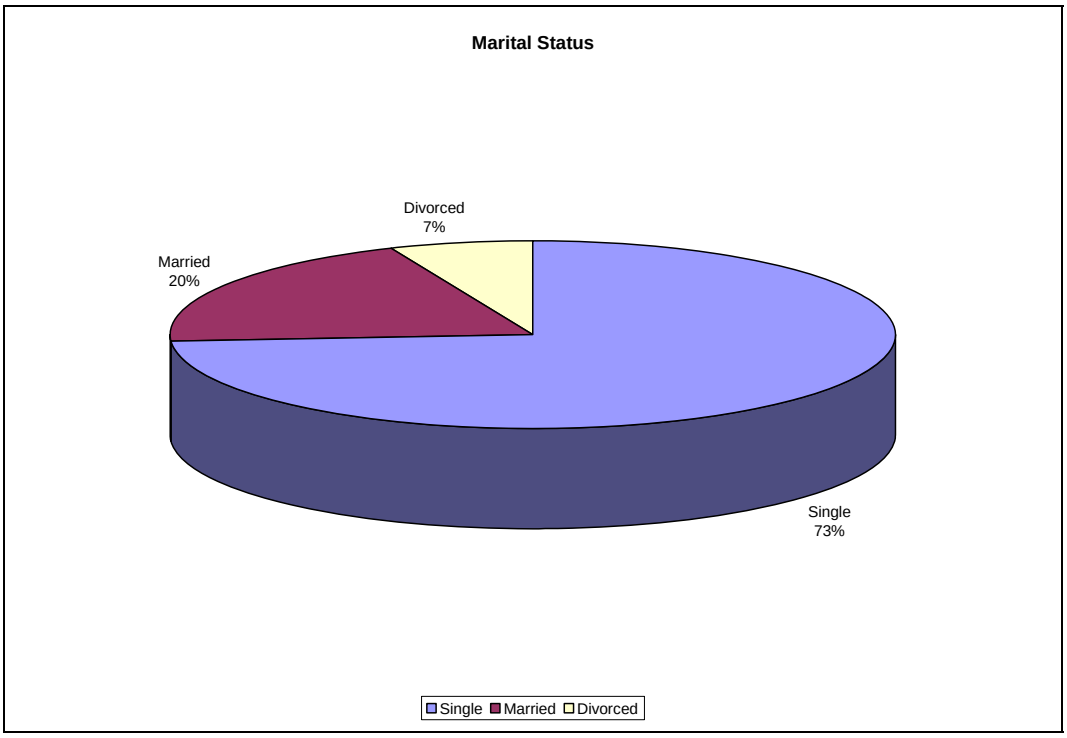


Figure 6: Marital Status of Survey Participants

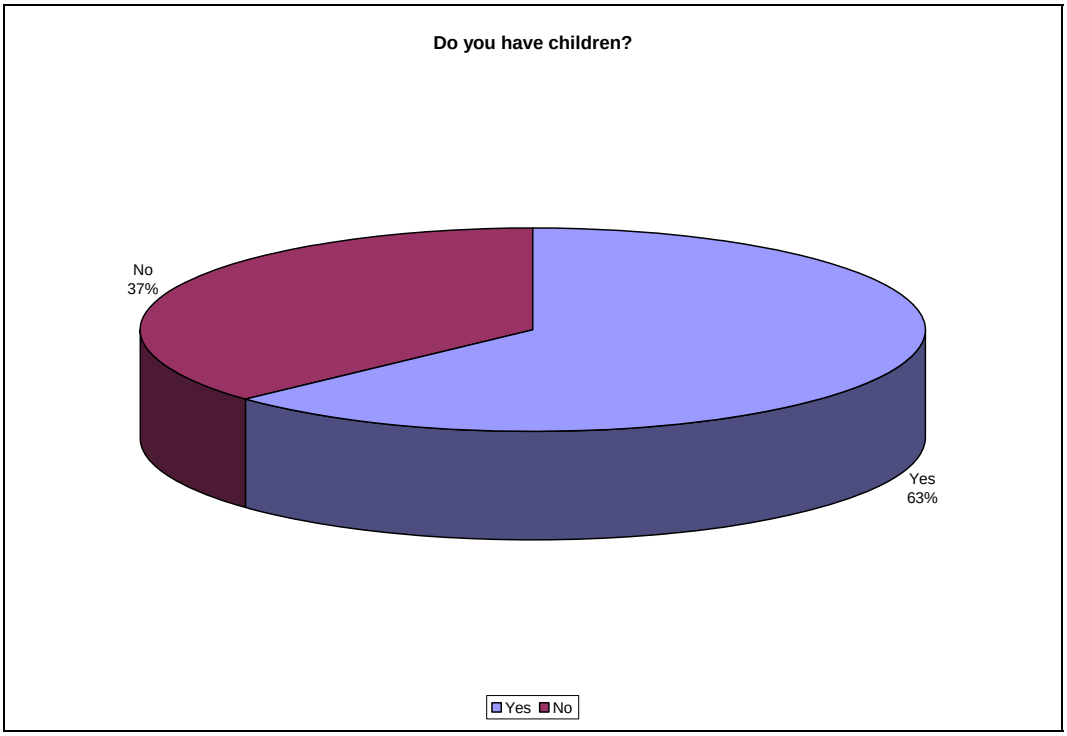


Figure 7: Percentage of Participants with Children

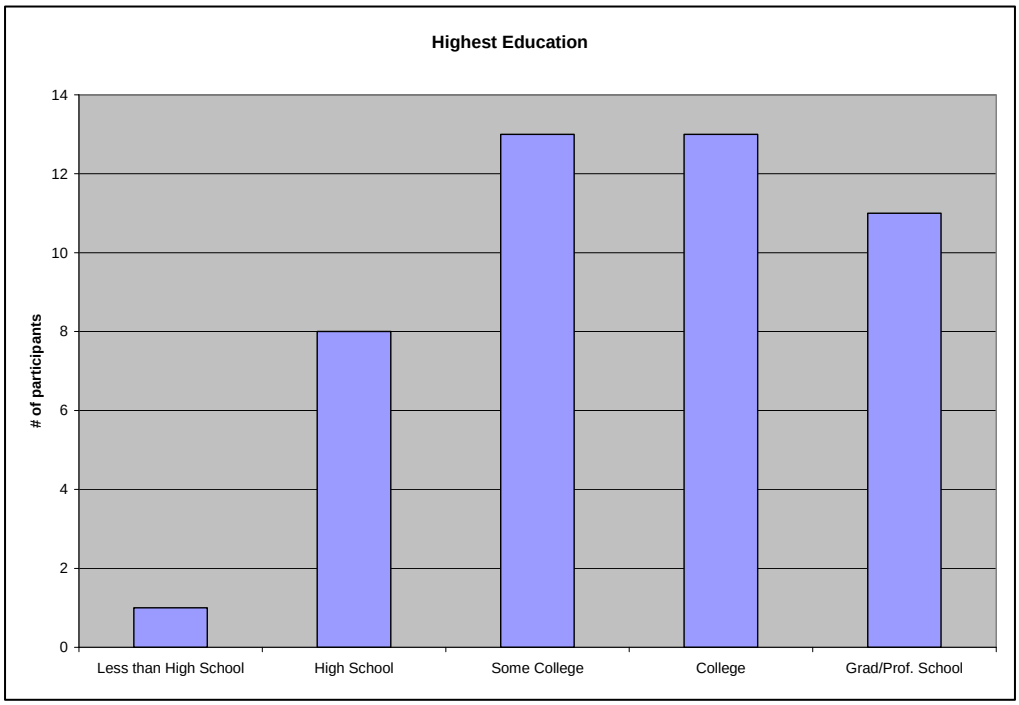


Figure 8: Highest Level of Education Completed by Survey Participants

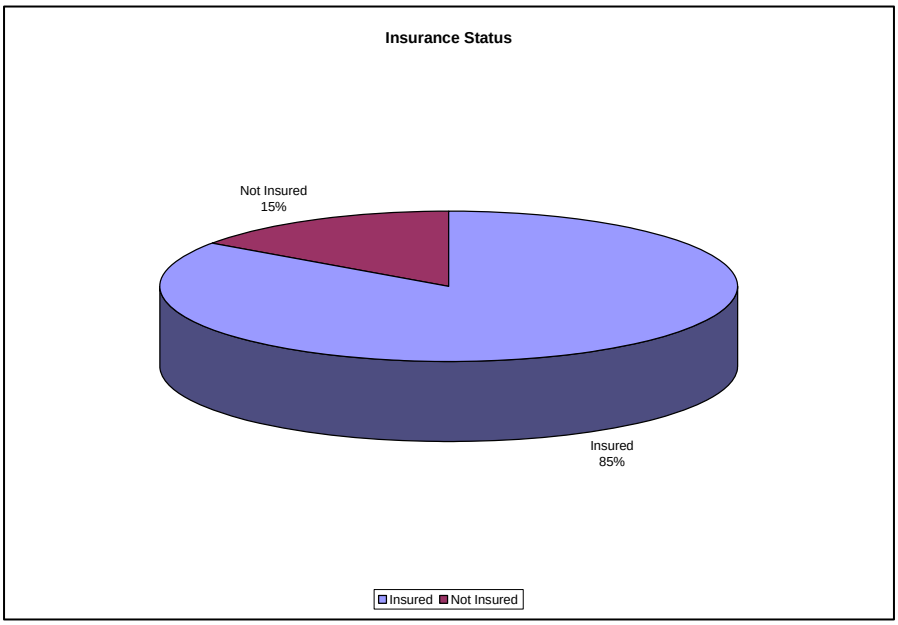


Figure 9: Insurance Status of Survey Participants

7.2 FEASIBILITY RESULTS

Of the 50 participants who were consented for the study, three were lost to follow-up during the process between pre-education and post-education. A total of 47 participants completed the pre to post questionnaire process with a 94% retention rate. Thirty-seven of the 50 participants who were consented completed the whole process from the pre-questionnaire to the 1 month follow-up questionnaire. The result was a 74% retention rate through the whole process and a 79% retention rate from the completion of post questionnaire to 1 month follow-up questionnaire. The overall conclusion of the retention rate was that there were a significant number of participants that were retained throughout the study compared to other studies of its kind. [8; 9; 64]

The continued participation of the two locations for outreach Ms. Ida's Epiphany and A Second Glance Salon and Spa also made this outreach project feasible. Both shops continued to participate over the five month period with each holding a Sickle Cell Trait Testing Day at their shop.

7.3 IMPACT RESULTS

7.3.1 Acquisition and Knowledge Retention Results

Pre vs. Post Questionnaire

A total of nine questions were used to evaluate acquisition and retention of knowledge about sickle cell disease and sickle cell trait education given to each study participant. Seven of the nine questions analyzed were found to have significant ($p<0.05$) gain of knowledge (Table 1).

The two statements that were not found to be significant included "African Americans are at higher risk for sickle cell disease." and "Sickle cell disease can be caught just like the common cold." The majority of participants knew these answers prior to education with 90.00% correct, 8.00% uncertain, and 2.00% incorrect for first statement and 82.00% correct and 16.33% uncertain for the second. These findings explain that the majority of participants knew that African Americans are at a high risk for sickle cell disease and that sickle cell disease is not contagious.

Each of the other seven questions were found to have significant knowledge gain ($p<0.05$). (See Table 1) This shows a knowledge gain of the etiology and inheritance of sickle cell disease and an improvement in the understanding of sickle cell trait. All of the knowledge questions were also grouped together to analyze overall knowledge gain. The results from the paired t-test showed that there was significant overall knowledge gain from the pre-questionnaire to the post-questionnaire with a p-value of <0.001. The mean score after education for the post questionnaire was 8.26 out of 9 compared to a score of 5.6 out of 9 before education. The result was an average score of 92% after education from 62% before education. (See Figures 8 and 9)

Pre vs. 1 Month Follow-up Questionnaire

The same nine knowledge questions were evaluated for impact from the pre-questionnaire to the 1 month follow-up questionnaire. Using the Fisher's Exact Test five of the nine questions resulted in significant knowledge gain through the complete study process as shown in Table 1. Significance was based on a p-value of <0.05. These five statement were "Sickle cell disease can affect the red blood cells", "Sickle cell disease can cause pain and strokes", "Genetic carriers will not develop symptoms of sickle cell disease", "If only one parent is a carrier of sickle cell

trait, they have no chance of having a baby with sickle cell disease", and "Typically, both parents of a child need to be carriers of sickle cell trait in order to have a child with sickle cell disease." Overall there was knowledge gain from the pre questionnaire before education to the 1 month follow-up questionnaire Overall participants knew from the beginning to end that sickle cell disease is not contagious and that it is more common in African Americans, and number of participants had forgotten that children with sickle cell disease can have infections and pneumonia. A small number of participants also forgot that sickle cell disease is inherited. Another reason that the p-value was not found to be statistically significant is because the sample size in the Pre to 1 Month Follow-up category was lower than the Pre to Post category.

When looking at the overall knowledge gain of participants through the whole study there was a significant gain of knowledge. Overall knowledge gain was analyzed using a paired t-test with significance determined by a p-value of <0.05. When analyzed using the paired t-test the p-value for overall knowledge gain, the p-value was <0.001 showing significant knowledge gain of participants from the pre-questionnaire to the final questionnaire in the study. The mean scores after education was 8.27 out of 9 in the 1 month follow-up questionnaire compared to 5.65 out of 9 score before educations. Participants scored an average of 92% on the knowledge portion of the 1 month follow-up questionnaires compared to 63% before education.

Post vs. I Month Follow-up Questionnaire

The results of the knowledge gain from the post-questionnaire to the 1 month follow-up questionnaire did not show significant knowledge gain. The mean values from the two questionnaires were 8.26 and 8.27 out of 9.00. This was expected given that there was no additional education between the time of the post-questionnaire and the final questionnaire.

Table 1: Results of Knowledge Gain from Pre, Post, and 1 Month Follow-up Questionnaires

Knowledge Statements		Pre-Questionnaire Percent	Post-Questionnaire Percent	1 Month F/U Questionnaire Percent	Fisher's Exact Test Pre vs. Post	Fisher's Exact Test Pre vs. 1 Month F/U
Sickle cell disease affects the red blood cells.	**Agree:**	70.00	100.00	94.59	**<0.001**	**0.024**
	Uncertain:	24.00	0.00	2.70		
	Disagree:	6.00	0.00	2.70		
Children with sickle cell disease are at risk for infections and pneumonia.	**Agree:**	78.00	95.74	91.89	**0.027**	0.190
	Uncertain:	22.00	2.13	2.70		
	Disagree:	0.00	2.13	5.41		
Sickle cell disease can be caught just like a cold.	**Agree:**	0.00	4.26	2.70	0.091	0.056
	Uncertain:	16.00	0.00	0.00		
	Disagree:	82.00	95.74	97.30		
Sickle cell disease can cause pain and strokes.	**Agree:**	74.00	93.62	91.89	**0.021**	**0.035**
	Uncertain:	20.00	2.13	2.70		
	Disagree:	6.00	4.26	5.41		
African Americans are at a higher risk of being carriers of sickle cell trait.	**Agree:**	90.00	97.87	100.00	0.203	0.240
	Uncertain:	8.00	2.13	0.00		
	Disagree:	2.00	0.00	0.00		
Genetic carriers will not develop symptoms of sickle cell disease.	**Agree:**	26.00	65.22	75.68	**0.001**	**<0.001**
	Uncertain:	38.00	4.35	5.41		
	Disagree:	36.00	30.43	18.92		

Table 1 Continued

If only one parent is a carrier of sickle cell trait, they have no chance of having a baby with sickle cell disease.	**Agree:**	22.00	85.11	83.78	**<0.001**	**<0.001**
	Uncertain:	20.00	0.00	2.70		
	Disagree:	58.00	14.89	13.51		
Typically, both parents of a child need to be carriers of sickle cell trait in order to have a child with sickle cell disease.	**Agree:**	32.00	95.74	94.59	**<0.001**	**<0.001**
	Uncertain:	22.00	0.00	0.00		
	Disagree:	46.00	4.26	5.41		
Sickle cell disease is inherited.	**Agree:**	78.00	97.87	97.30	**0.015**	0.056
	Uncertain:	16.00	2.13	0.00		
	Disagree:	6.00	0.00	2.70		

7.3.2 Concern Results

There were no significant changes in concern from pre to post, pre to 1 month follow-up, or post to 1 month follow-up questionnaires. The majority of participants disagreed that sickle cell disease carrier testing was worrisome. A slight increase was observed in post and 1 month follow-up questionnaire categories of people agreeing and disagreeing that sickle cell trait testing was worrisome. This dual increase can mostly likely be attributed to participants being about to resolve their feeling of uncertainty about trait testing after education with 28.00% being

uncertain before testing to 10.64% and 8.11% uncertain in the post and 1 month follow-up questionnaire.

The overall results of concern using the paired t-test found that concern did not change through the course of the study. Most participants found that trait testing was not worrisome to them. In conclusion, they had a low level of concern about being worried about trait testing. (See Figure 8 and 9)

Table 2: Concern Results

Concern Statement		Pre-Questionnaire Percent	Post-Questionnaire Percent	1 Month F/U Questionnaire Percent	Fisher's Exact Test Pre vs. Post	Fisher's Exact Test Pre vs. 1 Month F/U
Sickle cell disease carrier testing worries me.	**Agree:**	8.00	12.77	16.22	0.259	0.315
	Uncertain:	28.00	10.64	8.11		
	Disagree:	64.00	76.60	75.68		

7.3.3 Attitude Results

A number of questions regarding attitude were considered significant from the pre to the post questionnaire. Using the Fisher's Exact Test significance was evaluated ($p<0.05$), there was a change in participants support for sickle cell disease carrier testing for communities, encouraging their partner to be tested for sickle cell trait if they were found to be a trait carrier, and understanding the role of a genetic counselor. Participants found that after education their outlook was more positive on carrier screening in communities ($p=0.015$). They also deciphered

the importance of trait testing for their partner if they were found to be a carrier ($p=0.012$). This identifies that an open communication for a discussion about sickle cell disease, the importance of knowing one's carrier status, and the value of family planning are important to participants. These findings suggest that attitude is good and that most people value the importance of carrier screening and family planning. The reason behind the Pre to 1 Month follow-up questionnaire not having the same significance in the Pre to Post questionnaires is most likely due to the difference in sample size between the two questionnaires. It is also because there was not a great change in responses of participants of this sample who completed the pre to 1 month follow up questionnaire. Their feelings about carrier testing for their partner and carrier testing for communities remained most commonly the same from pre to 1 month follow up questionnaire.

The other aspect of information that was gained from analysis of attitude questions was that people felt like they had a better understanding of the role of a genetic counselor after receiving education about sickle cell disease at the barbershop and beauty salon. This was also the most significant attitude finding when comparing the pre-questionnaires to the 1 month follow-up questionnaire ($p < 0.001$). Overall, participants had a better understanding of a genetic counselor by the completion of the study.

Table 3: Attitude Results

Attitude Statements		Pre-Questionnaire Percent	Post-Questionnaire Percent	1 Month F/U Questionnaire Percent	Fisher's Exact Test Pre vs. Post	Fisher's Exact Test Pre vs. 1 Month F/U
I support SCD carrier testing for communities.	**Agree:**	82.00	97.87	97.30	**0.015**	0.056
	Uncertain:	6.00	0.00	0.00		
	Disagree:	12.00	2.13	2.70		
I support SCD carrier testing and medical services for communities in barbershops and beauty salons.	**Agree:**	84.00	91.49	97.30	0.355	0.199
	Uncertain:	8.00	4.26	2.70		
	Disagree:	8.00	4.26	0.00		
I would encourage my partner to be tested for SCT if I was found to be a trait carrier.	**Agree:**	82.00	100.00	100.00	**0.012**	0.240
	Uncertain:	8.00	0.00	0.00		
	Disagree:	10.00	0.00	0.00		
I understand the role of a genetic counselor.	**Agree:**	56.00	93.62	94.59	**<0.001**	**<0.001**
	Uncertain:	40.00	4.26	2.70		
	Disagree:	4.00	2.13	2.70		
I feel like meeting with a genetic counselor is helpful to my understanding of SCD and SCT testing.	**Agree:**	78.00	93.62	97.30	0.070	0.056
	Uncertain:	18.00	2.13	0.00		
	Disagree:	4.00	4.26	2.70		
Genetic counseling is	**Agree:**	90.00	100.00	100.00	0.056	0.115

Table 3 Continued

an effective way to learn, understand, get resources, and support about genetic conditions.	**Uncertain:**	10.00	0.00	0.00		
	Disagree:	0.00	0.00	0.00		

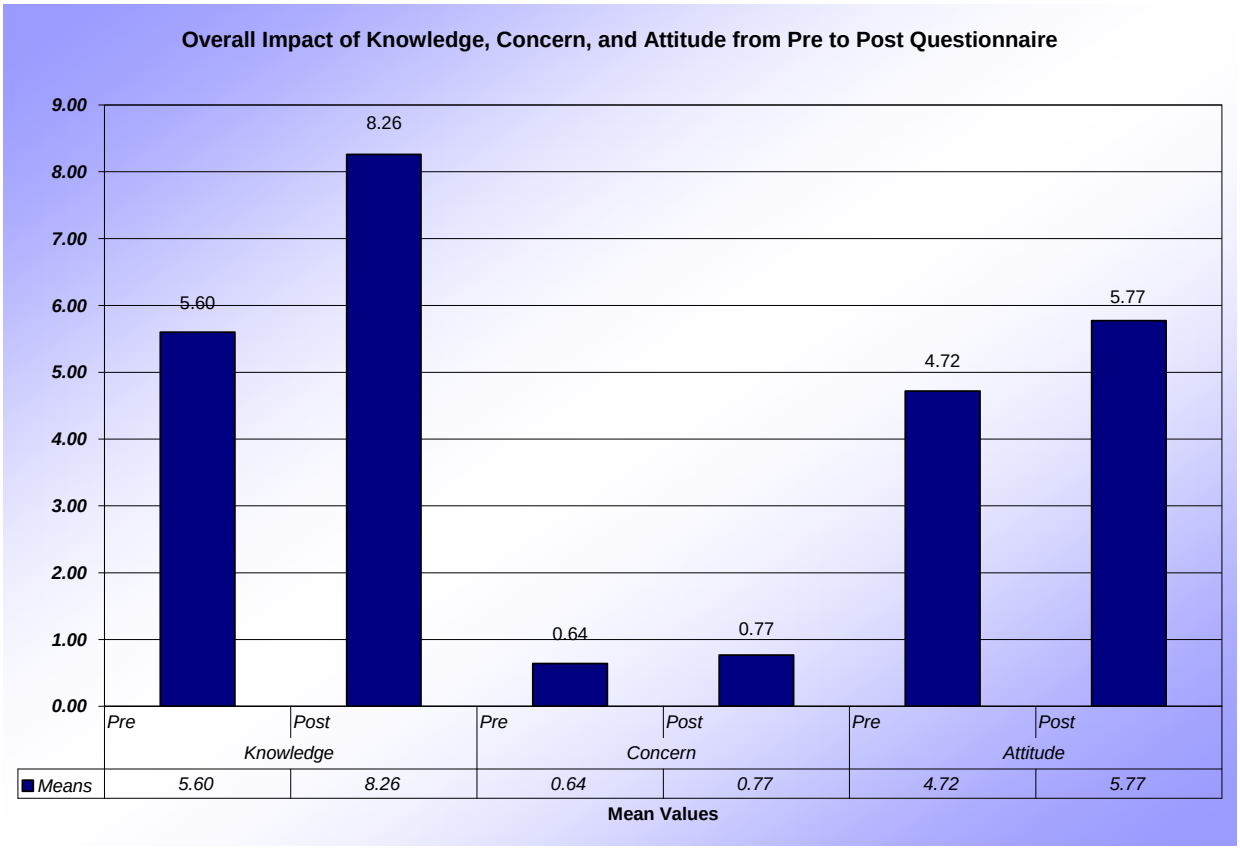


Figure 10: Overall Impact of Knowledge, Concern, and Attitude from Pre to Post Questionnaire

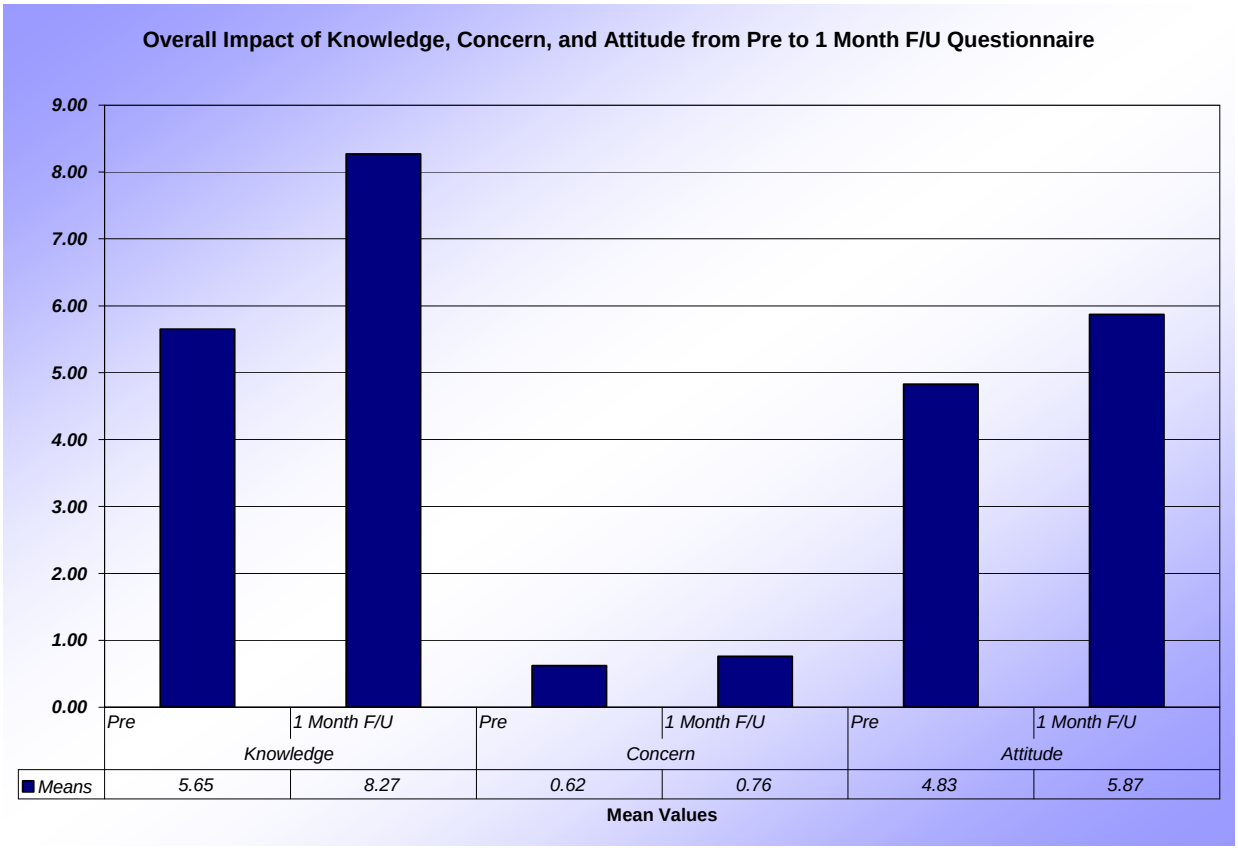


Figure 11: Overall Impact of Knowledge, Concern, and Attitude from Pre to 1 Month Follow-up Questionnaire

7.3.4 Qualitative Analysis

From the six open ended questions, a number of themes were observed within the data. Overall participants felt that they had a positive attitude about how they felt about receiving health services from their local barbershop/beauty salon. A total of 91% of participant felt positively about receiving health services at the shops during the post questionnaire and 97% felt positively about receiving health services at the shops during the one month follow-up questionnaire.

Responses pertaining to if genetic counseling was a helpful experience, found that 91% of participants' experiences with genetic counseling were helpful and 97% expressed that it was

a positive experience in the 1 month follow-up questionnaire. A number of participates wrote detailed responses on their thoughts of genetic counseling. One participant said, "It enlightened me on misconception about sickle cell disease." Two others commented, "I learned something I didn't know" and "It gave me answers to unanswered questions." Most participants had some amount of exposure to sickle cell disease. Further education at the barbershop and beauty salon, gave additional information to participants about sickle cell disease and sickle cell trait that they may not have become exposed to in the future.

When asked about if participants felt like they learned something, 91% and 100% of participants responded that they did learn something from the education during the post questionnaire and 1 month follow-up questionnaires. This extended to the majority of participants feeling that they could share what they had learned about sickle cell disease and sickle cell trait with others. In the post questionnaire, 90% of participants said that they felt like they could share the information they learned with others. When asked in the 1 month follow-up questionnaire 95% of participants felt that they could share the information with others. In the post questionnaire no information was gathered from 4 participants. After a month of retaining information more participants were prone to share the information with others in the future.

Looking at recommendations of participants to a program like this to others, participants responded in the post questionnaire that 96% would recommend a program like this to other and 100% of participants would recommend a program like this to others in the 1 month follow-up questionnaire. There overall feeling suggested that they felt positively about the study and its aims. Participants identified the benefit of doing education in the community and would recommend it to others. No information was gathered from four participants pertaining to if they would recommend this program to others. That is how the 96% rate was gathered.

8.0 DISCUSSION

8.1.1 Specific Aim 1: Feasibility

Overall this study has shown that community outreach in the barbershop or beauty salon is a feasible method of providing genetic education, genetic counseling, and genetic testing regarding sickle cell disease and sickle cell trait. The setting of the barbershop or beauty salon was beneficial for recruiting participants. The locations provided a steady number and turn around of people. Within a 2 ½ month period, 50 participants were consented for the study. Loss to follow-up occurred between pre to post questionnaire administration. At times, participants would complete the pre-questionnaire and receive education then get their haircut and comeback to finish the post questionnaire. Three participants were lost to follow-up in this process because they left without completing the post questionnaire or they completed the pre-questionnaire and did not comeback to go through the education and post questionnaire. If they had not received education, then there was no attempt to re-contact the participant. If they did receive education, then the participant was contacted to avoid loss to follow-up.

Loss to follow-up also occurred between the post to 1 month follow-up questionnaire. All participants were called up to four times for the final 1 month questionnaire. If there was no contact by the fourth attempt, then questionnaires were mailed with a self addressed stamped envelope. Of the 47 participants that completed the post-questionnaire at the barbershop or

beauty salon 10 were lost to follow-up. A total of 37 participants completed the study with a 74% retention rate that was consistent with other studies assessing feasibility. Incentive may be an additional option in order to improve loss to follow-up. Incentives whether monetary or gift oriented may motivate participants to complete the entire process of the study increasing the retention rate and improving feasibility and impact.

The majority of people at the barbershop and beauty salon were open to education at the shops. Some people declined participation in the study with a number of people receiving just education. This is one area that could be explored more deeply by examining the reasons and emotions behind refraining from participating in research. Exploring the differences between the demographics of participants and looking at other factors such as motivation can help tailor future study approaches to community outreach in the barbershop or beauty salon setting in order to better reach underserved groups. Overall, participants and those that received education were enthusiastic about receiving education at their barbershop or salon. Many expressed that it was a good to come into the community to do education on sickle cell disease and sickle cell trait. The majority of participants were comfortable with the idea of receiving education at the location. Some even referred the study PI to their community leaders to organize sickle cell disease and sickle cell trait education for other organizations. Feasibility of collecting participants, retaining them through the study, and attaining their feedback about their thoughts about genetic education, genetic counseling and genetic testing were all feasible in the study.

Providing the education in the open setting of the waiting area also exposed those that were coming to the two locations who the PI may not have been talking to specifically to the information. A number of people saw other people receiving the education and completing the questionnaires and were interested in the information themselves and wanted to participate in the

study. Keeping the information open to discussion helped facilitate education as a conversation with the community. At times, a group of people would engage in the educational discussion. This was also found in the 1 month follow-up surveys in participants' responses to feeling comfortable about talking to others about sickle cell disease and sickle cell trait. Therefore not only was education feasible in the shops, but those that received education felt that they were able to go out and continue to talk about sickle cell disease and trait with others in their community.

Participants also showed a positive attitude about genetic counseling and its feasibility in the barbershop or beauty salon. Only 78% of participants knew the role of a genetic counselor before education. The majority of participants did not know the role or were uncertain. By the end of the study, 94% of the participants responded that they did understand the role of a genetic counselor and that they thought that genetic counseling is helpful. This change can help facilitate participant's comfort level with medical professionals when they see them working in their community.

Reviewing responses on genetic testing and screening in the community, both were considered positive by participants. Looking at participants who completed the entire study, 76% did not know their trait status before the receiving education. After the study, 82% were interested in knowing their sickle cell trait status. This suggests the feasibility of providing trait testing at the shops. This was attempted as a separate outreach project from the study providing free of charge trait testing to the community. Given the proper conditions for blood draw, trait testing could be offered and preformed at the shops. The ability to provide outreach in a setting that is comfortable and trusted by the target community can improve the feasibility and success of a community outreach project.

The final aspect of feasibility and one of the most important was the retention of the participating locations. Both shops were extremely cooperative throughout the study. Arrangements were made for educational/recruitment days as well as trait testing days separate from this study. Involvement of the staff was also a key component to the feasibility of the study. Staff members were educated about the study, and a number of the staff participated in the study as well. Educating the staff about the study gave them the opportunity to talk with their clients about the education about sickle cell disease and sickle cell trait at the shop. Having the barber and stylists talk about sickle cell disease and sickle cell trait created the trust and by creating this more people were informed and therefore interested in the education.

8.1.2 Specific Aim 2: Impact

Overall this study has shown the impact of genetic education, genetic counseling, and genetic testing in barbershops and beauty salons in the African American community by testing the acquisition and retention of knowledge of sickle cell disease and sickle cell trait with the use of the study questionnaires. Looking at the three main categories of knowledge, concern, and attitude, both the knowledge and attitude categories illustrated a significant change in participants' responses following education. Participants showed a gain in overall knowledge of sickle cell disease and sickle cell trait. There was also increased knowledge of the role of genetic counselors and a change in participants' attitude toward sickle cell trait carrier testing for communities and talking to their partner about trait testing.

Knowledge was a main area of evaluation for understanding the impact of doing education at the barbershop and the beauty salon, and a significant amount of knowledge was gained with a p-value of 0.001. A brief individualized educational session utilizing visual aids,

slides, and take home material in the barbershop and beauty salon setting was given to each participant. Most participants had heard about sickle cell disease and sickle cell trait in the past and had some of knowledge of the condition. This previously known knowledge was often that sickle cell disease more commonly affects people of African descent and that sickle cell disease is not caught like the common cold. There was a significant gain of knowledge in the other questionnaire questions about sickle cell disease pertaining to what types of cells are affected, the natural history of the condition, and the inheritance of the sickle cell disease. The concept of inheritance and inheritance pattern was not something commonly known by the study participants. This is an area that could be looked at in closer detail. Greater education in the school systems about genetics and inheritance could be one area for further growth. Some participants had learned a little about inheritance, but were unsure if a person who was a carrier had a risk of having a child with sickle cell disease. Some participants who were older had no prior understanding to the inheritance pattern of the disease. Even after education, there were some participants who still did not understand this concept. Simplifying this concept as much as possible can facilitate a better understanding of this concept. Identifying those who are having trouble with the concept is important in order to simplify the concept and take further measures to explain it with further additional visual aids or drawing out the inheritance. In this instance, the main concept of having an increased risk for sickle cell disease when both parents are carriers of sickle cell trait is the most important concept to pass along to participants.

Concern about sickle cell trait testing in participants showed no significant change after educations. Participants remained unworried about sickle cell disease trait testing before and after testing. Seventy-eight percent of participants did not know their trait status even though they were not worried about trait testing. This statement may have been taken two different

ways by participants. Some may have thought of the question as, "Does trait testing worry me?" while others may have not been worried about trait testing because they did not know the implications of sickle cell disease, and they were not concerned by trait testing. After education they may have been more concerned, but because they were informed they responded the same way because they were not fearful of getting testing; therefore, not worried by it. Further study and research about the topic of concern could shine light on the reasoning behind level of concern among participants. More questions pertaining to concern could be beneficial in better understanding levels of concern among this population.

More investigations could be used to better understand concern about trait testing and the public's motivations for getting testing. Twenty-six percent of participants that completed the whole study process had stated that they knew their trait status. If people are unconcerned, what is it that is keeping them from learning their status? Looking into motivations and perceptions about trait testing would provide better insight to this area. The barbershop and beauty salon location could be a good area to investigate this issue. Further study, could be carried looking at how many people after education went out and pursued trait testing. Comparing the number of people who pursued trait testing when they were referred to a clinic to when they had the option of receiving it at the barbershop or beauty salon.

In examining attitude, overall attitude remained positive over the course of the study. Most participants saw the benefit of community outreach. A number of participants commented that they wished that they had other medical topics that they could learn about this way. It would be beneficial for further outreach to be extended to other medical issues since feasibility in the community has been confirmed. When asked about how participants felt about receiving health services at their barbershop and beauty salon, the majority of participants responded positively to

the education. There seemed to be a good attitude toward education specifically medical education in the community. Participants even responded that they felt like they could go and talk to others about what they learned. The impact of this response is significant not only because the education impacted the individual educated, but it extended to others beyond those in the study.

Clearly, there was a significant amount of impact evaluated both quantitatively and qualitatively. Future studies can further extend the barbershop/beauty salon education model to include other health related outreach for medical issues such as heart disease or diabetes. The location can facilitate learning and education. It can also be a place of research. In terms furthering this study, looking at the motivations behind responses would help better understanding the target population and tailor future studies. Because this population has been underserved in the past, does not mean that it will be underserved in the future. Bringing the education to the people can increase education and increase motivation to act on the education. This can involve improving their own health and seeking their own healthcare through a short education session. With this, it is important to keep in mind the sensitivity of the subject. Not everyone is willing and open to education or learning about sickle cell disease or sickle cell trait. Utilizing sensitive education material tailored to the target population is vital. Moving forward, people can see what other people have done at their barbershop and can pursue their own interest in their health.

9.0 CONCLUSION

In conclusion, this study was successful at assessing both specific aims. Feasibility of delivering a health education module on genetic education, genetic counseling, and genetic testing in the community was successfully demonstrated. The impact of knowledge acquisition and retention of sickle cell disease and sickle cell trait education showed improvement in knowledge after education and retention of knowledge 1 month following education. Concern about trait testing remained the same throughout the study with the majority of participants not worried or indifferent about trait testing. Participants' attitudes about genetic education, genetic counseling, and trait testing also remained high. The most significant finding in the attitude category was a better understanding of the role of a genetic counselor. The overall results exhibited that the education at the barbershop and beauty salons had an impact on participants.

The success of this study was possible through the continued participation of all parties involved. The barbershop and salon were the integral part of this study, and without the help from the Center for Minority Health the strong relationship with the shops and the study would have never occurred. The two shops were always active and interested in participation and education. Barbers and stylists took an active role in educating themselves and their clients. Their participation facilitated the success of the project, and the number of participants interested in taking part in the study.

Future studies can use the information gained from this study to create new outreach projects not only for sickle cell disease, but for any other community based educational outreach. The results can help development health related education materials on genetic conditions common in the African American community. Medical and public health professionals can use the insights gained from this study to provide better education and outreach to the community through non-clinical settings in the community.

APPENDIX A: Approval Forms and Letters

University of Pittsburgh
Institutional Review Board

3500 Fifth Avenue
Pittsburgh, PA 15213
(412) 383-1480
(412) 383-1508 (fax)
http://www.irb.pitt.edu

Memorandum

To: CECILIA RAJAKARUNA

From: SUE BEERS, PhD, Vice Chair

Date: 10/6/2008

IRB#: PRO08020016

Subject: The Barber/Beauty Shop Visit…It's More Than Just Getting Your Hair Cut
Testing and Education of Sickle Cell Trait in Your Neighborhood Barber/Beauty Shop

Your research study has received expedited review and approval from the University of Pittsburgh Institutional Review Board under 45 CFR 46.110 (7).

Please note the following information:

Approval Date:	10/6/2008
Expiration Date:	10/5/2009

PLEASE NOTE THAT IT IS THE INVESTIGATOR'S RESPONSIBILITY TO REPORT TO THE IRB ANY UNANTICIPATED PROBLEMS INVOLVING RISKS TO SUBJECTS OR OTHERS [SEE 45 CFR 46.103(B)(5) AND 21 CFR 56.108(B)]. THE IRB REFERENCE MANUAL (CHAPTER 3, SECTION 3.3) DESCRIBES THE REPORTING REQUIREMENTS FOR UNANTICIPATED PROBLEMS WHICH INCLUDE, BUT ARE NOT LIMITED TO, ADVERSE EVENTS. IF YOU HAVE ANY QUESTIONS ABOUT THIS PROCESS, PLEASE CONTACT THE ADVERSE EVENTS

COORDINATOR AT 412-383-1480.

THE PROTOCOL AND CONSENT FORMS, ALONG WITH A BRIEF PROGRESS REPORT MUST BE RESUBMITTED AT LEAST ONE MONTH PRIOR TO THE RENEWAL DATE NOTED ABOVE AS REQUIRED BY FWA00006790 (UNIVERSITY OF PITTSBURGH), FWA00006735 (UNIVERSITY OF PITTSBURGH MEDICAL CENTER), FWA00000600 (CHILDREN'S HOSPITAL OF PITTSBURGH), FWA00003567 (MAGEE-WOMENS HEALTH CORPORATION), FWA00003338 (UNIVERSITY OF PITTSBURGH MEDICAL CENTER CANCER INSTITUTE).

Please be advised that your research study may be audited periodically by the University of Pittsburgh Research Conduct and Compliance Office.

Children's Hospital of Pittsburgh

3705 Fifth Avenue
Pittsburgh, PA 15213-2583

412-692-5055

PEDIATRIC HEMATOLOGY/ONCOLOGY AND BLOOD AND MARROW TRANSPLANTATION

Marty Ostrow Outpatient Clinic
4B385 DeSoto Wing
Fax: 412-692-6675

Administrative Suite
4B330 DeSoto Wing
Fax: 412- 692-7693

TO: Lakshmanan Krishnamurti, M.D., Director
Sickle Cell Program at Children's Hospital
Division of Hematology/Oncology,
Children's Hospital of Pittsburgh
University of Pittsburgh

FROM: A Second Glance Wellness and Spa
215 North Highland Ave., Pittsburgh, PA 15206
(Full name and address of facility)

RE: Permission of Distribute Information, Provide Education, Medical Services Relating to Trait Testing and Genetic Counseling, and Recruit Participants to the study.

DATE: 9/1/08

I understand that the Sickle Cell Program at Children's Hospital is looking to see the educational impact and feasibility of providing Sickle Cell Disease education in barbershops and beauty salons in the community. I understand that a genetic counseling student will offer the service education, trait testing, and genetic counseling in my shop to my clients.

Please accept my signature below as permission for The Sickle Cell Program at Children's Hospital to use this facility to provide the services listed above to the community.

Children's Hospital of Pittsburgh

3705 Fifth Avenue
Pittsburgh, PA 15213-2583

412-692-5055

PEDIATRIC HEMATOLOGY/ONCOLOGY AND BLOOD AND MARROW TRANSPLANTATION

Marty Ostrow Outpatient Clinic
4B385 DeSoto Wing
Fax: 412-692-6675

Administrative Suite
4B330 DeSoto Wing
Fax: 412- 692-7693

TO: Lakshmanan Krishnamurti, M.D., Director
Sickle Cell Program at Children's Hospital
Division of Hematology/Oncology,
Children's Hospital of Pittsburgh
University of Pittsburgh

FROM: Ms Ida's Epiphany Barber Shop
822 Wood St., Pittsburgh, PA 15221
(Full name and address of facility)

RE: Permission of Distribute Information, Provide Education, Medical Services Relating to Trait Testing and Genetic Counseling, and Recruit Participants to the study.

DATE: 9/1/08

I understand that the Sickle Cell Program at Children's Hospital is looking to see the educational impact and feasibility of providing Sickle Cell Disease education in barbershops and beauty salons in the community. I understand that a genetic counseling student will offer the service education, trait testing, and genetic counseling in my shop to my clients.

Please accept my signature below as permission for The Sickle Cell Program at Children's Hospital to use this facility to provide the services listed above to the community.

APPENDIX B: Pre and Post Questionnaires

PRE AND POST QUESTIONNAIRES

Sickle Cell Disease Educational Awareness
Pre-Education Questionnaire

Please check whether you agree, disagree, or are uncertain about the following statements.

	AGREE	UNCERTAIN	DISAGREE
Sickle cell disease affects the red blood cells.			
Children with sickle cell disease are at risk for infections and pneumonia.			
Sickle cell disease can be caught just like a cold.			
Sickle cell disease can cause pain and strokes.			
African Americans are at a higher risk of being genetic carriers of sickle cell disease.			
Genetic carriers will not develop symptoms of sickle cell disease.			
If only one parent is a carrier of sickle cell trait, they have no chance of having a baby with sickle cell disease.			
Typically, both parents of a child need to be a carrier of sickle cell trait in order to have a child with sickle cell disease.			
Sickle cell disease is inherited.			
Sickle cell disease carrier testing worries me.			
I support sickle cell disease carrier testing for communities.			
I support sickle cell disease carrier testing and medical services for communities in barber or beauty shops.			
I would encourage my partner to be tested for sickle cell trait if I was found to be a trait carrier.			
I understand the role of a genetic counselor.			
I feel like meeting with a genetic counselor is helpful to my understanding of sickle cell disease and sickle cell carrier testing.			
Genetic counseling is an effective way to learn, understand, get resources, and support about genetic conditions.			

Sickle Cell Disease Educational Awareness
Post-Education Questionnaire

Please check whether you agree, disagree, or are uncertain about the following statements.

	AGREE	UNCERTAIN	DISAGREE
Sickle cell disease affects the red blood cells.			
Children with sickle cell disease are at risk for infections and pneumonia.			
Sickle cell disease can be caught just like a cold.			
Sickle cell disease can cause pain and strokes.			
African Americans are at a higher risk of being genetic carriers of sickle cell disease.			
Genetic carriers will not develop symptoms of sickle cell disease.			
If only one parent is a carrier of sickle cell trait, they have no chance of having a baby with sickle cell disease.			
Typically, both parents of a child need to be a carrier of sickle cell trait in order to have a child with sickle cell disease.			
Sickle cell disease is inherited.			
Sickle cell disease carrier testing worries me.			
I support sickle cell disease carrier testing for communities.			
I support sickle cell disease carrier testing and medical services for communities in barber or beauty shops.			
I would encourage my partner to be tested for sickle cell trait if I was found to be a trait carrier.			
I understand the role of a genetic counselor.			
I feel like meeting with a genetic counselor is helpful to my understanding of sickle cell disease and sickle cell carrier testing.			
Genetic counseling is an effective way to learn, understand, get resources, and support about genetic conditions.			

How do you feel about receiving health services in your local barber/beauty shop?

_____

_____

Was genetic counseling a helpful experience for you and how?

_____

_____

Do you feel like you learned something from this program?

_____

_____

Do you feel like you can talk to others about what you have learned about sickle cell disease and trait testing?

_____

Would you recommend a program like this for others?

_____

_____

Demographic Data: Please circle the answer which best applies to you

Sex: Female Male

Age (Years): 16-20 21-25 26-30 31-35 36-40 41-45 46-50 50+

Highest Education: Less Than High School High School Graduate

Some College College Graduate Graduate/Professional School

Marital Status: Married Single Divorced

Number of Children: _____

Insurance Status: Insured Not Insured

Thank You for Your Participation and Help!!!

Sickle Cell Disease Educational Awareness
1 Month F/U-Education Questionnaire

Please check whether you agree, disagree, or are uncertain about the following statements.

	AGREE	UNCERTAIN	DISAGREE
Sickle cell disease affects the red blood cells.			
Children with sickle cell disease are at risk for infections and pneumonia.			
Sickle cell disease can be caught just like a cold.			
Sickle cell disease can cause pain and strokes.			
African Americans are at a higher risk of being genetic carriers of sickle cell disease.			
Genetic carriers will not develop symptoms of sickle cell disease.			
If only one parent is a carrier of sickle cell trait, they have no chance of having a baby with sickle cell disease.			
Typically, both parents of a child need to be a carrier of sickle cell trait in order to have a child with sickle cell disease.			
Sickle cell disease is inherited.			
Sickle cell disease carrier testing worries me.			
I support sickle cell disease carrier testing for communities.			
I support sickle cell disease carrier testing and medical services for communities in barber or beauty shops.			
I would encourage my partner to be tested for sickle cell trait if I was found to be a trait carrier.			
I understand the role of a genetic counselor.			
I feel like meeting with a genetic counselor is helpful to my understanding of sickle cell disease and sickle cell carrier testing.			
Genetic counseling is an effective way to learn, understand, get resources, and support about genetic conditions.			

How do you feel about receiving health services in your local barber/beauty shop?

_____

_____

Was genetic counseling a helpful experience for you and how?

_____

_____

Do you feel like you learned something from this program?

_____

_____

Do you feel like you can talk to others about what you have learned about sickle cell disease and trait testing?

_____

Would you recommend a program like this for others?

_____

_____

Did you know your trait status before this being educated?

_____

Do you want to know your trait status after having education?

_____

APPENDIX C: Educational Script

EDUCATIONAL SCRIPT

1. Introduction:

    A. Introduce myself and that I am a graduate student working at the Sickle Cell Program at Children's Hospital working on a study looking at the benefits of providing education about sickle cell disease in the community.

    B. Ask if individual would mind talking with me for a few minutes.

    C. Explain that we are talking to people in the community about sickle cell disease to see what they already know about the disease and educate about the disease. We are also interested in their feelings about sickle cell trait testing, doing community outreach at the barbershop/ beauty salon, and how they feel about genetic counseling.

    D. Ask if they are over 18 years of age and interested in learning about sickle cell disease and sickle cell trait.

        - If yes, then ask if they are interested in participating in taking a pre and post questionnaire, receiving education about sickle cell disease and sickle cell trait, and taking a one month follow-up questionnaire.

- If no, ask if they are interested in the education, learning about sickle cell disease and sickle cell trait and provide handouts.

E. If yes, then review consent form with participant.

F. Give coded pre-questionnaire.

2. Education

A. Use Slides (Figure

B. Begin with a conversation of what sickle cell means to the individual. Do they know anyone who has sickle cell disease? What do they know about sickle cell disease?

C. (Slide 1) Sickle cell disease is an inherited blood disorder that affects the red blood cells in our body causing them to change their shape.

D. Normal red blood cells are soft and carry oxygen throughout the body. Sickle red blood cells are hard and sticky and do not carry oxygen as well throughout the body.

E. It can be very painful and can cause various medical complications.

F. The reason that we come to do education about it at the shops is because sickle cell disease and sickle cell trait most commonly affects people of African American descent.

a. 1 in 500 African Americans affected with sickle cell disease

b. 1 in 12 African Americans affected with sickle cell trait

c. Ask if they know the difference between sickle cell disease and sickle cell trait. Explain that we have who of every gene in our body. One we get from mom and one we get from dad. Someone with sickle cell disease has

two non-working copies of the sickle cell gene. Someone who has sickle cell trait has one non-working copy of the gene. These individuals are carriers of sickle cell trait. People with sickle cell trait have no symptoms of sickle cell disease and will not develop sickle cell disease. Those with sickle cell disease have severe medical complication.

G. (Slide 2) Review features of sickle cell disease touch on each point also mentioning that we can see strokes in some children with sickle cell disease as well.

H. (Slide 3) Review inheritance

   a. Sickle cell disease cannot be caught like the common cold it is not contagious. In order for someone to have sickle cell disease, both of their parents would need to be carriers of sickle cell trait.

I. (Slide 4) Reiterate inheritance points

   a. One some with sickle cell trait is healthy and not going to develop sickle cell disease or symptoms of the disease.

   b. Typically, both parents of a child with sickle cell disease need to be carriers of sickle cell trait in order be at risk for having a child with sickle cell disease.

   c. If only one parent is a carrier of sickle cell trait, they have no chance of having a child with sickle cell disease

J. (Slide 5) Discuss who at risk for sickle cell trait.

   a. Anyone is at risk, but it is more common in African Americans.

b. Variants of sickle cell disease can be found in people of Mediterranean, Middle Eastern, Indian, Asian, and Hispanic populations.

K. (Slide 6) How can you tell if you have trait?

a. The only way to tell is to be tested with a simple blood test.

b. Since 1992, Allegheny County has mandated sickle cell diseases screening on their newborn screening panel, but those born before then do not know their trait status from newborn screening.

c. We will be offering free of charge trait testing for individual's who would like to learn their trait status. We will be offer testing at the shop with the sickle cell disease program physician, phlebotomist, and sickle cell disease educator.

L. (Slide 7) Importance of knowing trait status

a. The most important reason to be aware of your trait status is to find out if you could possibility have a baby with sickle cell disease. The only way to find out is for you and your partner to be tested.

M. (Slide 8) If you find out you do have sickle cell trait

a. If you have trait remember this is not going to cause you to be sick or have sickle cell disease. People who have trait are healthy.

b. This is useful information that helps you prepare for children that could possibly be at risk for having sickle cell disease.

c. Genetic counselors are available to help explain the disease, the risks, discuss testing, and other options with you.

d. The most important thing you can do is be responsible for the best health care for your child, and one way to do that is to be informed about your trait status.

APPENDIX D: Educational Slides and Handouts

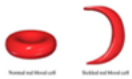
EDUCATIONAL SLIDES AND HANDOUTS

Sickle Cell Disease

What does it mean to you?

What is Sickle Cell Disease?

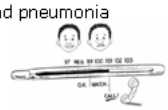
- Sickle cell disease is an inherited disease that affects the red blood cells in our body causing them to change their shape.



- It is can be a very painful disease.
- It has the highest occurrence amongst those with African American decent.
  - 1 in 500 African Americans are affected with sickle cell disease.
  - 1 in 12 African Americans has sickle cell trait.

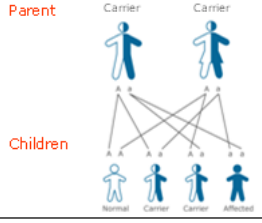
Features of Sickle Cell Disease

- Fever over 101°C
- Increased risk for infections and pneumonia
- Chest pain
- Difficulty breathing
- Getting tired very easily
- Abdominal swelling
- Headache that is different than usual
- Sudden weakness or loss of feeling
- Severe pain that just won't go away
- Sudden change in vision



How does someone get sickle cell disease?

- Sickle cell disease is caused when each parent passes a non-working copy of the hemoglobin gene to their child.
- It **cannot** be caught like a cold.

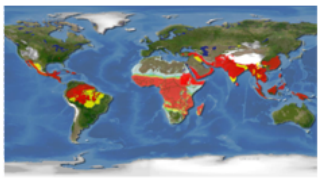


More information about inheritance

- Genetic trait carriers will not develop symptoms of sickle cell disease.
- If only one parent is a carrier of sickle cell trait, they have no chance of having a baby with sickle cell disease.
- Typically, both parents of a child need to be a carrier of sickle cell trait in order to have a child with sickle cell disease.

Who is at risk for **sickle cell trait**?

- Anyone can carry sickle cell trait; however, it is most common in **African Americans**.
- It also can be found in people of Mediterranean, Middle Eastern, Indian, Asian, and Hispanic backgrounds.



How can you tell if you have trait?

- The only way to tell is to be tested with a simple blood test.
- Since 1992, Allegheny County has offered sickle cell disease screening on their newborn screening panel, but those born before then do not know their testing status.
- We will be offering free of charge trait testing for individual's who would like to learn their trait status.

Why is it important to know your trait status?

- The most important reason to be aware of your trait status is to find out if you could possibly have a baby with sickle cell disease. The only way to find out is for you and your partner to be tested.

What can you do if you have trait.

- If you have trait remember this is **not** going to cause you to be sick or have sickle cell disease.
- People who have trait are **healthy**.
- **This is useful information** that helps you prepare for children that could possibly be at risk for sickle cell disease.
- Genetic counselors are also available to help explain the risks and discuss prenatal testing, and other options with you.
- The most important thing you can do is be responsible for the best health care for your child.

Thank you!

Where should I go with more questions?

Doctors, nurses and genetic counselors at Children's Hospital of Pittsburgh are available to answer your questions.

For More Information and Testing

- Children's Hospital of Pittsburgh
  Division of Hematology
  412-692-6059
- Children's Sickle Cell Foundation Inc.
  www.cscfkids.org
  412-537-8973
  Gove Business Center
  226 Paul St.
  Pittsburgh, PA 15211
- Magee-Womens Hospital Medical Genetics
  412-641-4168 *or*
  1-800-454-8155
- Sickle Cell Disease Association of America
  www.sicklecelldisease.org
- Sickle Cell Society Inc.
  412-371-0628
  7643 Frankstown Ave
  Pittsburgh, PA 15208

Children's
Hospital of Pittsburgh

www.chp.edu

cscf | Children's Sickle Cell Foundation, Inc.

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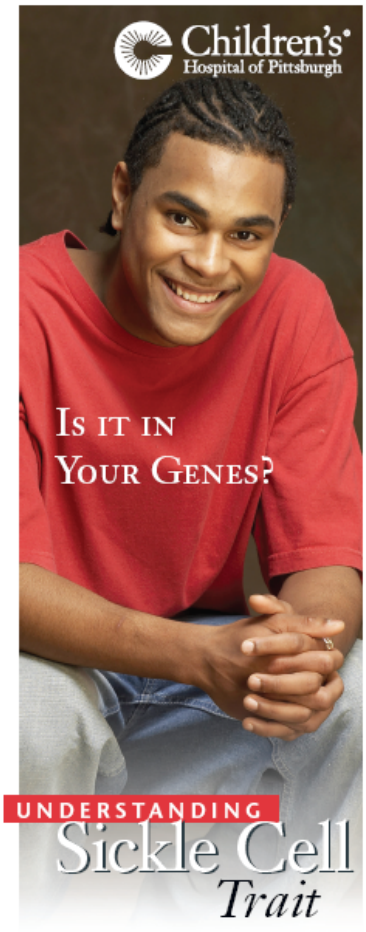


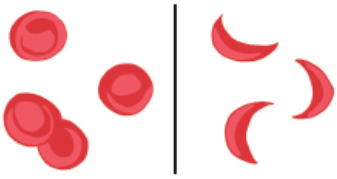
UNDERSTANDING Sickle Cell *Trait*

UNDERSTANDING

Sickle Cell *Trait*

What Is Sickle Cell Disease?

Sickle Cell Disease is a serious disease of the red blood cells that causes the cells to change their shape. It can be a very painful disease. Sickle cell disease is caused when each of your parents pass you a non working gene, before birth. You can not catch it from being around someone with sickle cell disease.



Normal Red Blood Cells

Sickled Red Blood Cells

What Is a Gene?

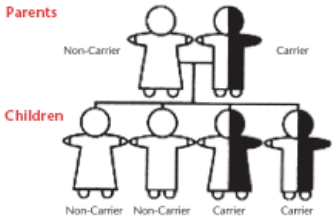
Genes are the instructions that parents pass to their children for different traits such as eye color or diseases such as Diabetes. We have two copies of every gene in our body. We receive one copy from our mother and one from our father before birth. There is no way to control which genes are passed from parents to children. The sickle gene (S) affects the red blood cells.

What Is Sickle Cell Trait?

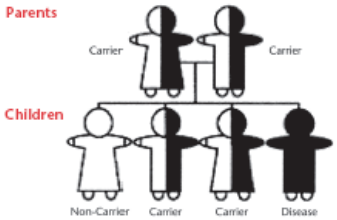
Sickle Cell Trait occurs when a person inherits a working gene (A) and a non working Sickle gene (S), they are also called a "carrier". Sickle cell trait is not a disease. People with sickle cell trait cannot tell they have it without being tested. If two people who have sickle cell trait have a baby, that child is at risk to receive a non-working sickle (S) gene from each parent, which will cause sickle cell disease.

The most important reason to be aware of sickle cell trait is to find out if you can possibly have a baby with sickle cell disease. The only way to find out is for you and your partner to get tested. The different genes for the disease come from both parents- not just one.

When one parent is a carrier



When both parents are carriers



Who is at risk for Sickle Cell Trait?

Anyone can carry a sickle cell trait; however, some populations are at a higher risk than others are. Sickle cell trait is most common in African Americans. However, people of Mediterranean, Middle Eastern, Indian, Asian and Hispanic background are also at an increased risk to have sickle cell trait so many people should be tested.

How can you tell if someone has Sickle Cell Trait?

The only way to tell of someone has sickle cell trait is to be tested. Some blood tests are more useful than others. The best tests give detailed results. They can find the (S) gene. But they can also find OTHER genes, such as the (C) gene or the Beta thalassemia gene which can combine with the (S) gene to cause other forms of sickle cell disease.

What can I do if I have Sickle Cell Trait?

If you find out that you have Sickle Cell Trait, your spouse or partner should also be tested. Knowing ahead of time that your future children may be at risk gives you time to prepare for and learn more about caring for a child with a serious illness. Genetic counselors are also available to help explain the risks and discuss prenatal testing, and other options. The most important thing you can do is be responsible for the best health care for your child.

APPENDIX E: Advertisements

ADVERTISEMENTS FOR TRAIT DAYS

Figure 12: Advertising for Ms. Ida's Epiphany Barbershop

Figure 13: Advertisement for A Second Glance Beauty Salon

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