A MODEL FOR THE IMPLEMENTION OF SICKLE CELL DISEASE AND TRAIT EDUCATION INTO PUBLIC SCHOOL HEALTH CLASSES AT THE MIDDLE SCHOOL LEVEL

by

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Diana Young, M.S.

Since September 1992, Sickle Cell Trait (SCT) has been a standard on the state of Pennsylvania's newborn screen. Therefore, as of 2006, all middle school aged children will have had newborn screening. Currently, no information about Sickle Cell Disease or trait is included in the curriculum of the Pittsburgh Public Schools (K-12).

There are three goals for this project: To educate the teenagers of Pennsylvania about Sickle Cell Disease and Trait, teaching them what their trait or disease status means for their health, and for their future children's health; To provide the schools with an appropriate curriculum and; To create a module that can be repeated in school systems throughout the country.

Approval of funding by the Pittsburgh Public Schools Board of Education allowed for the hiring of three teachers to aid in the writing of an appropriate curriculum. The curriculum includes three lessons that involve watching a video, writing informational materials, learning about genetics, and creating a poster.

To assess this program and its effectiveness, health teachers critiqued the curriculum designed by their peers and provided feedback on ways to improve the implementation of new health material to existing lesson plans. After making modifications from this feedback, future projects may include adding a Sickle Cell Disease and Trait curriculum into the high school and

eventually elementary schools. We would like to eventually share the curriculum online through a dependable source so schools throughout the nation have access to the lesson plans.

The project is relevant to the field of public health because it directly educates individuals about SCT and SCD at an early age. They will be able to reference this information in the future when they are faced with making reproductive decisions.

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PREFACE

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1.0 INTRODUCTION

Since September of 1992 the Commonwealth of Pennsylvania has been screening for the hemoglobinopathies, including Sickle Cell Disease (SCD) and Sickle Cell Trait (SCT). ("National Newborn Screening and Genetics Resource Center: Current Newborn Screening (NBS) Conditions by State " 2007) This screening is a result of the publication of the NIH consensus statement that mandated the implementation of newborn screening for the hemoglobinopathies which was adopted in 1987. ("Consensus conference. Newborn screening for sickle cell disease and other hemoglobinopathies," 1987) Studies show that children with SCD have an increased susceptibility to bacterial infections, especially pneumococcal sepsis. If penicillin is taken daily starting in the first few months of life, the risk of infection and potential morbidity and mortality is greatly reduced. These studies suggest the value of screening in the neonatal period. (Gaston & al., 1986)

The primary goal of screening for the hemoglobinopathies during the newborn period is to detect children affected with SCD, begin prophylactic penicillin, and introduce them to more attentive pediatric care. However, Hemoglobin electrophoresis is the most often used screening technique and many SCT carriers are identified. The Children's Hospital of Pittsburgh (CHP) is contracted by the Commonwealth of Pennsylvania to notify and follow-up any abnormal hemoglobinopathy results from the newborn screen for 32 counties in Western Pennsylvania. Each year 12-15 newborns are diagnosed with SCD in CHP catchments area, and are evaluated at Children's Hospital of Pittsburgh (Kladny, Gettig, & Krishnamurti, 2005). Treatment begins immediately. Approximately 600 newborns are identified on the newborn screen to carry SCT each year; however, no official form of follow-up is required by the state. (Kladny et al., 2005)

Since the screening began in 1992, many of these children are presently approaching high school age, and are uninformed about SCT, what it is, or if they are carriers. This also means that these children are approaching child bearing age, but may not have information to enable informed decisions based on their trait status. Many of the children identified on the newborn screen as SCT carriers are now attending schools that are a part of the Pittsburgh Public School system, a system in which the majority of the students are of an African American ancestry. No curriculum on SCD or SCT currently exists in the public school system. The Pennsylvania Standards for Health, Safety, and Physical Education states, "Pennsylvania's public schools shall teach, challenge and support every student to realize his or her maximum potential and to acquire the knowledge and skills need to: Analyze how personal choice, disease and genetics can impact health maintenance and prevention. (10.1.9.E)." (Academic Standards for Health, Safety, and Physical Eduation, 2002) Keeping these standards in mind, and knowing the population that is attending Pittsburgh Public Schools, it was proposed to develop a curriculum to implement into middle school classes. This project aims to create an appropriate curriculum that can be maintained in the school system for years to come.

2.0 SPECIFIC AIMS

2.1 SPECIFIC AIM I

Providing access to new educational materials :

Specific Aim: To create a curriculum with information about Sickle Cell Trait and Sickle Cell Disease to supplement the existing health curriculum in middle school classes.

Hypothesis: Not all schools have the time, money, or resources to create a new curriculum for their health classes. A new curriculum can provide variety and valuable information for teachers and students alike.

Plan: Join with the Pittsburgh Public Schools and several of their teachers to create a curriculum appropriate for middle school health classes.

2.2 SPECIFIC AIM II

Knowledge of Sickle Cell Disease and Sickle Cell Trait:

Specific Aim: To educate the youth of Pittsburgh by teaching them what their trait or disease status means for their health, and for their future children's health, as they are approaching a reproductive age; To educate the teachers and students of the Pittsburgh Public School system about SCD.

Hypothesis: The lack of knowledge about SCT status among Pittsburgh youth may be due to the lack of knowledge about SCD as a serious disease and its pattern of inheritance. The importance of knowing trait status may be emphasized and improved by teaching youth about the disease and the genetics of the disease.

Plan: Utilizing the curriculum created in conjunction with the teachers of the Pittsburgh Public School, information about Sickle Cell Trait and Sickle Cell Disease can be taught to the majority of middle school-aged youth in Pittsburgh. The effectiveness of this teaching method will be evaluated by using individual teacher surveys, as well as qualitative assessments answered by the teachers. Changes will be made where necessary.

3.0 BACKGROUND AND SIGNIFICANCE

3.1 SICKLE CELL DISEASE

Sickle Cell Disease (SCD) is an inherited disease that affects the body's red blood cells. It is inherited in an autosomal recessive manner. Hemoglobin is the main substance in a red blood cell that helps the red blood cells to carry oxygen throughout the body. β -globin is a major subunit of hemoglobin. Mutations in the β -globin gene cause a different type of hemoglobin to be produced. If each of an individual's two β -globin genes has a mutation, this person is considered to have Sickle Cell Disease. The two abnormal β -globin units cause a change in the shape of the red blood cells from being round and soft to being hard and sickle shaped (Figure 1). (Wethers, 2000b)



Figure 1: Healthy and sickled red blood cells (http://www.defiers.com/scd.html, 3/5/2007)

The cells' change in shape causes them to become stiff, which can lead to vascular occlusion. This process involves the sickled shaped cells becoming 'stuck' in blood vessels and preventing oxygen from traveling to tissues and organs resulting in pain, tissue ischemia, and potential organ damage depending on the tissue that is being affected.(Wilson, Krishnamurti, & Kamat, 2003) For example, if a block occurs in the brain, this can lead to a stroke. It is thought that a red blood cell not only changes its shape to become sickled, but it also expresses more adhesion molecules on its surface. This extra adhesion, or 'stickiness', may be the initiating factor for vasoocclusion. (Frenette, 2002)

A sickled red blood cell also loses its ability to effectively transport oxygen throughout the body. This results in a shorter life span for the red blood cells (about 20 days, compared to the usual 120 days), which leads to the anemia that has helped give Sickle Cell Disease the more commonly recognized name of Sickle Cell Anemia. As mentioned, SCD is inherited in an autosomal recessive manner. If an individual is homozygous for a mutation in each of their two β -globin genes, they are considered to have SCD. Most red blood cells contain hemoglobin A, which is functional and capable of carrying oxygen all over the body. Mutations cause hemoglobin to change from being a healthy type of hemoglobin, called hemoglobin A (HbA), to a less functional hemoglobin S (HbS) molecule. There are other mutations that can occur in the β -globin genes that can cause a person to be a compound heterozygote and be affected with SCD. Other examples of genotypes of individuals affected with SCD are a person with hemoglobin S and one of the following: hemoglobin C, β thalassemia (HbS β^+ and HbS β^0). (Wilson et al., 2003) There is much variability in the presentation of the disease regardless of genotype, but in general, individuals with HbSS or HbS β^0 -thalasemmia are thought to have a more severe presentation. (Wilson et al., 2003) (Miller et al., 2000)

3.1.1. Incidence of Sickle Cell Disease

Sickle cell disease is the most common single gene disorders in individuals of African American descent. About one in 375 people are born with SCD. (Wethers, 1027) This equates to about 2,000 children born with SCD in the United States per year. Due to the autosomal recessive nature of the disease, SCT is more common than SCD and affects approximately 8-10%, or approximately 1 in 10 to 1 in 12 African Americans. (Shafer et al., 1996) (E. Vichinsky, Hurst, Earles, Kleman, & Lubin, 1988) The majority of the time, individuals are carrying hemoglobin S trait, with hemoglobin C trait being the next most common mutation. (Wethers, 2000a) While SCD is the most common single gene disorder in African Americans, it is also common in individuals of Mediterranean, Middle Eastern, Indian, Arabian, and South and Central American descent. (Wethers, 2000a)

Sickle Cell Disease demonstrates a theory called heterozygote advantage. Individuals with SCT appear to display a resistance to fatality due to malaria that homozygotes do not have.(Aidoo et al., 2002) This is thought to be the reason why SCT and thus SCD are more common in areas where malaria is common. These areas include Central Africa, the Middle East, and India, and are illustrated in Figures 2 and 3.



Figure 2: Areas affected by malaria ("Distribution of malaria,")



Figure 3: Areas showing a higher incidence of Sickle Cell Trait ("Percent of the population that

has the sickle cell allele,")

Heterozygotes are also known as individuals with SCT, or trait carriers. These individuals have more than 50% normal hemoglobin and generally will not show signs associated with SCD. However, in some unusual cases, they may show signs such as anemia. (Wethers, 2000a) Individuals with SCT do have the ability to pass their abnormal hemoglobin gene on to their offspring. Each child they have, has a 50% chance to inherit that abnormal hemoglobin gene. It is important for someone with SCT to be aware of their carrier status because if their partner has SCT as well, they have a 25% chance to have a child with SCD with each pregnancy. There is also a 50% chance to have a child with SCT, and a 25% chance to have a child without SCT or SCD with each pregnancy. (Karnon et al., 2000)

3.1.2 Natural History of Sickle Cell Disease

Sickle Cell Disease was first described by James B. Herrick and his intern, Ernest E. Irons in 1910. (Savitt & Goldberg, 1989) They noted anemia in man of West Indian descent with oddly shaped red blood cells. (Savitt & Goldberg, 1989) Since this first description, the understanding of SCD has increased drastically, not only in the description of its characteristics and the underlying pathophysiology, but also in the treatment of the disease. The characteristic features of SCD are typically a result of hemolysis or episodes of vascular occlusion that lead to either tissue ischemia or organ damage. Consequences of hemolysis may include anemia, jaundice, aplastic crisis, and gallstones. Vascular occlusion can affect all organs of the body, but is most likely to affect the lungs, spleen, and brain. (E. P. Vichinsky, 1991) (Elliott Vichinsky, 2002)

The most common, yet unpredictable characteristic is an acute vaso-occlusive event, or a hallmark pain episode. (Wilson et al., 2003) Pain crises are the most common cause of morbidity at any age. The earliest signs of a pain crisis can be seen in infants before the age of one year. The most prominent sign is called dactylitis, or "hand-foot syndrome". (Wilson et al., 2003) This painful swelling of the bones in the hands and/or feet (seen in Figure 4) may be a predictor of severe disease in a child. (Claster & Vichinsky, 2003)



Figure 4: "Hand-Foot Syndrome"

(<u>http://www.acta-clinica.kbsm.hr/Volume 39 2000/11/11.htm</u>) ("Voluminous hand in psoriatic and rheumatoid arthritis and algodystrophic syndrome,")

Acute chest syndrome (ACS) is the next most common cause of hospital admission, and the leading cause of mortality in patients with SCD. (Claster & Vichinsky, 2003) The best way to diagnose ACS in an individual is by identifying new infiltrate on a chest radiograph. Other symptoms involved in the diagnosis include fever, cough and chest pain. (Wilson et al., 2003) ACS may develop soon after a pain crisis, or it can also arise with an acute infection, most often a pulmonary infection. (Wilson et al., 2003)

Splenic sequestration is recognized by an enlarging spleen and a drop in hemoglobin levels, and is a result of immune dysfunction and an increase of infections. This most often occurs in children under the age of three. (Claster & Vichinsky, 2003) (Wilson et al., 2003) If untreated, a patient may progress to shock and eventually death. (Claster & Vichinsky, 2003)

Children with SCD have an increased risk for stroke, with approximately 10% of children with SCD having a stroke between the ages of four and six. (Wilson et al., 2003) Symptoms can present as hemiparesis, headaches, seizures, or changes in mental status. Strokes are a direct result of vascular occlusion occurring within the blood vessels of the brain, which can limit oxygen levels. (Wilson et al., 2003) (Wethers, 2000a) Some small strokes may go unnoticed, however, these "silent infarcts" have been shown to produce cognitive defects, and can increase risk for future strokes, and should be treated. (Claster & Vichinsky, 2003)

Another complication associated with vasoocclusion and SCD is priapism, or a prolonged painful erection in males. (Fowler, Koshy, Strub, & Chinn, 1991) Episodes usually last anywhere from 1 hour to more than 24 hours, and if prolonged priapism goes untreated it may lead to impotence. (Fowler et al., 1991)

A more serious complication associated with SCD is an aplastic crisis, or a temporary cessation of bone marrow activity, usually resultant of an exacerbation of an individual's anemia. (Wilson et al., 2003) An infection is usually what triggers the anemia that leads to an aplastic crisis, with the most common cause of infection being human parvovirus. (Wilson et al., 2003)

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Most individuals have enough hematocrit to recover spontaneously, however some people with low hematocrit levels may need a red blood cell transfusion. (Wilson et al., 2003)

It may be easy to observe the physical characteristics associated with SCD, but many health care providers have noticed the psychological impacts that the disease can have on an individual, especially an adolescent. Adolescents with SCD can be behind in development by up to two years, including short stature and delayed sexual development. (Wethers, 2000a) (Wilson et al., 2003) Their growth will eventually reach an average size. (Wethers, 2000a) There are also issues the children deal with such as having to miss school or social functions due to an illness, which may make them feel isolated from their peers. (Wethers, 2000a) Another issue that individuals with SCD deal with is their ability to get fatigued faster than someone without SCD. They can participate in sports and other activities, but must realize that staying hydrated is important, as well as resting when they begin to feel too fatigued. (Wethers, 2000a) It is important to realize that SCD is a lifelong disorder, and not only affects children and their school work, but also adults and their careers, as well as many other aspects of life including, but not limited to peer and family relationships, and spirituality. (Ballas, 2002) Individuals with SCD will rarely be limited in their career choices, and should be encouraged to not let SCD take control of their lives. (Wethers, 2000a) Genetic counseling may be recommended to young adults who are planning to have a family. The inheritance of SCD and how it can relate personally to their reproductive future may prove a valuable piece of information for them. (Wethers, 2000a)

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3.1.3 Treatment of Sickle Cell Disease

Newborn screening has resulted in the ability of early detection of babies with SCD. This allows the early implementation of penicillin and folic acid being taken on a daily basis. Prophylactic penicillin was introduced as a standard of care to prevent infections, especially of pneumococcal sepsis. Since the introduction of prophylactic penicillin in 1986, the incidence of infection was reduced by 84%. (Elliott, Morgan, Day, Mollerup, & Wang, 2001; Gaston & al., 1986) Survival and life expectancy of children with SCD have also improved, demonstrating the importance of early detection on the newborn screen. (Wilson et al., 2003) Folic acid is taken daily to reduce the risk of bone marrow failure due to their chronic hemolysis. (Wilson et al., 2003) Because the risk of pneumococcal sepsis is so high in children with SCD, it is also standard of practice for these children to receive the 7-valent pneumococcal conjugated vaccine in infancy, or a similar 23-valent vaccine in childhood. Flu vaccines are recommended yearly, as well as a meningococcal vaccine for children who have shown splenic dysfunction. (Claster & Vichinsky, 2003) (Wethers, 2000a) The daily dosage of folic acid is to ensure an adequate amount for high erythrocyte turnover. (Wethers, 2000a)

Individuals with SCD disease will also need to establish routine care, not only with their PCP, but also with a specialty clinic or comprehensive care center. Being seen by physicians every six months allows for both physical examinations and the comparison of laboratory work for that specific patient. These regular check ups help the physicians to note any changes, and better adjust the care for each individual patient. (Wilson et al., 2003) These appointments also provide an opportunity to educate families about SCD and SCT, and their inheritance patterns.

Pain is the most common symptom associated with SCD, and therefore is consistently trying to be suppressed or controlled by patients, their families, and physicians. Treatment focuses on individual assessment and personalized therapy including relaxation and other home remedies. (Ballas, 2002) Treatment by physicians for a more severe pain crisis can include nonopioid and opioid analgesics such as morphine. During a hospitalization, emphasis is placed on hydration and managing any infections that may lead to future crises. (Ballas, 2002) Researchers have noted that patients with a high percentage of fetal hemoglobin (hemoglobin F) in their blood have a reduced amount of pain crises. (Platt et al., 1994) Further research has focused on keeping hemoglobin F levels elevated in patients with SCD. Recent use of hydroxyurea has been effectively shown to induce expression of hemoglobin F, and therefore reduce the incidence of pain crises. (Ballas, 2002; DeSimone et al., 2002)

Fevers can be a sign of infection and should be paid appropriate attention. Often times a chest x-ray will be done to rule out the risk of acute chest syndrome. Antibiotics will be given to control the infection, and hydration is key. (Wilson et al., 2003) Aplastic crisis and splenic sequestration are also associated with a high fever. (Wilson et al., 2003) Approximately one third of adults with SCD will die from either acute chest syndrome or stroke, so it is important for physicians to manage these two characteristics aggressively. (Platt et al., 1994) Acute chest syndrome needs to be treated quickly to prevent infection or more serious complications such as pulmonary failure or even death. Treatment can include oxygenation, hydration, antibiotics, and blood transfusions. (Wilson et al., 2003) Another potentially morbid characteristic of SCD is stroke. Strokes may present with hemiparesis, cranial nerve palsy, severe headaches, stupor, or coma. (Wilson et al., 2003) It has been found that screening at risk children with a transcranial Doppler ultrasound and following up with a blood transfusion greatly reduces the incidence of stroke. (Claster & Vichinsky, 2003)

Blood transfusions are used to treat several symptoms associated with SCD. This type of therapy increases the oxygen carrying capacity of blood by increasing the concentration of hemoglobin and decreasing the concentration of sickle hemoglobin. (Claster & Vichinsky, 2003) Straight transfusion is used when patients have a low hemoglobin concentration, while exchange transfusion should be used when the hemoglobin concentration is high so as not to increase viscosity. (Claster & Vichinsky, 2003) Indications for the need of a transfusion include acute chest syndrome, heart failure, multiorgan failure, stroke, splenic sequestration, and aplastic crisis. (Claster & Vichinsky, 2003) Transfusions have been shown to create a 90% reduction in risk for a future stroke. (Thompson & Thompson, 2006) It should not be used as treatment for steady state anemia, uncomplicated pain episodes, or minor surgery, as this may result in iron overload. (Claster & Vichinsky, 2003)

Sickle Cell Disease is not a curable disease in all patients; however, new research is ongoing regarding bone marrow transplantation (BMT). BMT can be curative for SCD. There are risks and morbidity associated with this procedure, so it is not available for all patients. The majority of research has been done on patients with a sibling-matched donor. To date, there has been a reported 94% survival rate, and an 84% cure rate for a bone marrow transplant (with a sibling-matched donor). (Walters et al., 2000) In patients with successful transplants there was noted to be normal erythropoiesis, improved growth, stable CNS findings, and stable pulmonary function in the majority. (Walters et al., 2000) Further research is being done in hopes of making this a cure available to all patients.

3.1.4 Molecular Genetics of Sickle Cell Disease

A healthy adult hemoglobin (hemoglobin A) molecule is made of two α globin chains and two β globin chains that bind oxygen in the red blood cells for transportation throughout the body. Hemoglobin S, or sickle hemoglobin results from a single point mutation of glutamic acid to valine. This is due to a substitution at the second nucleotide in the sixth codon in one of the two β globin chains. (Steinberg & Brugnara, 2003) The presence of value on this altered β globin chain causes it to polymerize with other molecules resulting in abnormally shaped red blood cells that are brittle and have difficulties traveling through blood vessels. (Steinberg & Brugnara, 2003) Not only does the shape of the red blood cell change, but it also impacts the membrane of the red blood cell. This mutation causes cellular dehydration, oxidative damage, and increased adherence of the red blood cells. (Frenette, 2002; Gladwin & Rodgers, 2000; Hebbel, 2000) The changes in cell shape and membrane properties are two key components for vaso-occlusion (figure 5). While SCD may be a monogenic disorder, it is multigenic in its presentation, indicating potential for secondary genetic factors. These other factors may play roles in increased production of cytokines, altered blood flow, coagulation abnormalities, and abnormal vascular regulation. (Nagel, 2001)



Figure 5: Binding and Polymerization of Hemoglobin S Molecules (Steinberg 1999)

Polymerization is also affected by several different factors, including deoxygenation, increased hemoglobin concentration, dehydration, acidity, temperature, and the presence of other hemoglobins, (for example, fetal hemoglobin inhibits polymerization). (Ballas, 2002) Polymerization is the even that gives red blood cells their characteristic shape in SCD. (Lonergan, Cline, & Abbondanzo, 2001) This event may be reversible with oxygenation, however, potential side effects may result in further dehydration of the red blood cell, adding to the sickling cycle. (Ballas, 2002; Lonergan et al., 2001) The constant cycle of polymerizationdepolymerization contributes to an increase in hemolysis and decreases the lifespan of a red blood cell from approximately 120 days to about 20 days. (Wilson et al., 2003)

3.2 THE PITTSBURGH PUBLIC SCHOOL SYSTEM

3.2.1 Background and Population

Public education officially began in Pittsburgh in 1835, and the school district was eventually formed in 1911 by a merger of the city's ward schools. (Isler et al., 2006) From 1911 until 1976 the board was comprised of 15 members. Beginning in 1976 the selection of the board moved to an election process that still exists today where 9 individuals are elected to represent 9 proportionate districts. (Isler et al., 2006)

Today, the Pittsburgh Public School system provides public education for the cities of Pittsburgh and Mount Oliver, Pennsylvania. According to the 2000 census, these two cities cover an area of 55.3 square miles, with a population of 342, 503. (Isler et al., 2006)In total there are 65 schools that provide education for 29, 445 students. (Smydo, 2006) These schools are staffed by 5,180 full time employees. At the middle school level, there are 4,001 students attending 11

different schools. (Isler et al., 2006) In addition to the 11 middle schools, there are also 19 schools that teach grades kindergarten through 8. Statistics based on ethnicity show that almost 61% of enrolled students have an African American background. (Isler et al., 2006)

3.2.2 School Curriculum

In the past few years, schools across the nation have been seeing reduced graduation rates including Pennsylvania, with a graduation rate of 80-89%. (Services, 2006) Reductions in the class sizes of Pittsburgh schools of 5.5% from fall 2005 to fall 2006 have been seen as families continue to move from the district and enroll their children in private schools and surrounding school districts, such as Woodland Hills. (Smydo, 2006) This drop marks at least the eighth consecutive year that the schools have seen a decline in enrollment. (Smydo, 2006) Speculations for the drop in enrollment include a decline in occupancy of public housing communities that surround the schools, an increase in discipline problems causing children to be removed from the school, dissatisfaction with the new combination of several elementary and middle schools, and finally parental dissatisfaction with academic achievements of their children in the schools. This final reason being the most prominent or at least the most controversial issue. (Smydo, 2006)

With these declines and the dissatisfaction of parents and students in mind, the superintendent of the schools has begun working toward academic revitalization. Last year, eight of the poorest performing elementary and middle schools were transformed into Accelerated Learning Academies (ALAs) as approved by the Board of Education on February 28, 2006. (Schools, 2006) The ALAs run on a longer school year by adding 10 extra days, as well as adding 45 extra minutes to each day for more instruction time. ("Pittsburgh Public Schools Accelerated Learning Academies Frequently Asked Questions," 2006) To gain extra help with

their curriculum content, the superintendent decided to outsource and use the *America's Choice* Design Model, which is developed by the National Center on Education and the Economy. ("Pittsburgh Public Schools Accelerated Learning Academies Frequently Asked Questions," 2006) This curriculum provides a routine and sets expectations for the students. This design encourages frequent monitoring, as to better individualize instruction to meet students' specific needs. ("Pittsburgh Public Schools Accelerated Learning Academies Frequently Asked Questions," 2006) This style of education may be leading the way for the rest of the Pittsburgh Public Schools.

To help produce a more solid core curriculum for other schools grades 6-12, the district contracted Kaplan K12 Learning Services. Kaplan, Inc. is an education company based out of New York City that provides learning and test preparation materials for all different grade levels. (Kaplan, 2001) The new core curriculums are being phased in over the next 2 years, and include subjects such as English, math, science and social studies. (Services, 2006) These core curriculums have a goal of preparing students for life after high school, and are based on the belief that a curriculum is not a textbook, but a program for learning. (Services, 2006) They use the curriculums as a 'map' for teachers to guide their students through the learning process. Their curriculums also organize learning expectations and align them to the districts resources and goals. ("Pittsburgh Public Schools Parent Information Sessions," 2006)

3.2.3 Teachers

To be a teacher in grades kindergarten through 12th in the Commonwealth of Pennsylvania, three main criteria must be met while applying for certification and a future teaching position. The first criteria item is the completion of a state-approved teacher education program (this can include either a baccalaureate or more advanced degree). This progarm must include a student teaching or intern experience. ("The Pennsylvania Bulletin: Rules and Regulations," 1999) The second qualification item is completion of Praxis I and Praxis II assessments with an appropriate qualifying score. ("The Pennsylvania Bulletin: Rules and Regulations," 1999) The Praxis I assessment is a pre-professional skills assessment (PPST) that is designed to measure basic skills such as reading, writing, and mathematics. ("The PRAXIS Series," 2007) The Praxis II assessment measures knowledge in specific subjects that the future teachers will be educating students about, as well as testing on general knowledge and teaching skills. ("The PRAXIS Series," 2007) The Pennsylvania Department of Education recognizes Praxis I and II test scores only taken in the past five years. ("The PRAXIS Series," 2007) The final step to obtaining certification in Pennsylvania is that all application materials must document the above mentioned criteria and the dates on which they were met. ("The Pennsylvania Bulletin: Rules and Regulations," 1999)

3.3 EDUCATION AND CURRICULUM IN MIDDLE SCHOOLS

Teacher qualifications vary on a state by state basis. Generally, the educational background remains consistent for most states, but the differences usually depend upon which PRAXIS tests are necessary for a certification. ("The PRAXIS Series," 2007) A national survey of teachers revealed that a good health teacher also has other qualities that don't just make them a teacher, but a valued health teacher. These qualities included their professional preparation along with a use of a variety of instructional techniques, skills in working with parents, knowledge of health content, and ability to work with community agencies/organizations. (Birch et al., 2001)

Implementing innovative instructional techniques or new curricula has proven a difficult and unsuccessful task for many teachers in the past. Previous studies have shown that between 33% and 85% of teachers do not use the entire curriculum created for a new topic, causing teachers to skip key materials to the curriculum. (Ringwalt et al., 2004) The suggested solution to this problem has been the creation of an adaptable curriculum with optional content, or one which teachers can adjust to fit their own style and schedule, as well as the needs of the students'. (Ringwalt et al., 2004) Another effective method in introducing new curriculum is teacher education prior to the implementation of the curriculum. Teacher knowledge about specific subjects has shown significant increase from before to after an in-service training day. (Levenson-Gingiss & Hamilton, 1989) Not only can an extensive in-service training increase a teacher's comfort level with the subject, but it can also increase their awareness so that they realize the importance or relevance of the topic. (Levenson-Gingiss & Hamilton, 1989) There are no specific reviews that cover the implementation of a SCD curriculum into the schools.

Tools for implementing a curriculum on genetic diseases into middle school classes are difficult to find. One woman, Debra Collins, noticed this deficit, and began a website designed to provide resources for individuals who teach human genetics for different grade levels. Highlighted throughout the web page and the links provided are the key points to cover when teaching about a genetic disease. These include inheritance pattern, clinical description, treatment, and testing or detection methods. (Collins) There are also several lesson plans included with suggestions for new and unique ways to teach. Several ideas involve the creation of an informational pamphlet about the specific disease. One lesson plan takes this idea a step further and has high school students using a pamphlet or poster they created to teach younger students. (Collins, April 12, 2007) Other teaching techniques included using books or games to teach inheritance patterns, with a specific idea of utilizing different colored candy to emphasize different genes. (Collins) Many lesson plans have a laboratory component to aid in the teaching of specific testing techniques for the diagnosis of a specific disease. If financial supplies are limited, there is a possibility to teach these lessons through a computer module or pictures. (Collins) Finally, a teaching method that helps students to personalize and further understand a disease is to use stories and personal accounts of individuals who are actually affected with the disease. (Collins) Another benefit of this specific website is that many resources are provided for genetic conditions in general, but also for particular diseases such as SCD. There is information appropriate for children of many ages, physicians, adults, as well as affected and unaffected individuals. (Collins)

4.0 MATERIALS AND METHODS

4.1 PARTNERSHIP

Members of the Children's Hospital Sickle Cell Clinic community and outreach education team attend health fairs and other community events. It was at one of these health fairs that a contact was formed with a member of the Pittsburgh Board of Education. Sarah Martin is the head of Health and Physical Education in the Curriculum and Instruction department of the Pittsburgh Public Schools. She noted the importance of SCT and SCD education and how it was lacking in the Pittsburgh Public Schools. It was proposed to this her that they create a curriculum together with Children's Hospital of Pittsburgh and the Pittsburgh Public Schools. Schools that were to be targeted first were the middle schools. This would allow the curriculum to be adjusted in the future for either a younger (elementary) or older (high school) student population. Children's Hospital of Pittsburgh (CHP) would provide factual information, while teachers from the school district would provide the knowledge in curriculum creation.

4.2 SPECIFIC AIM I: PROVIDING ACCESS TO NEW EDUCATIONAL MATERIALS

In order to hire teachers from the public school system to assist with the writing of the curriculum, the Board of Education would have to accept the proposed idea of hiring teachers to help write a curriculum on SCT and SCD, as well as the gift of money to fund the interested teachers. In December 2005, authorization to hire three middle school teachers at a rate of \$22.71 per hour was sought by the Pittsburgh Board of Education members at their monthly meeting. The official request stated the desire for CHP to pay three to four middle school teachers a regular workshop rate of \$22.71 per hour to aid in the development of supplemental curriculum materials relating to SCT and SCD. The Board of Education's resolution was to thank Children's Hospital of Pittsburgh, and accepted the action requested. (The action and resolution can be found in Appendix A)

In January 2006, after the winter break for the Pittsburgh Public Schools, a posting was given to human resources to hire three to four middle school teachers to aid in writing the curriculum. Over the next month, three teachers applied, and all three were interviewed. An example resume for a writer is found in Appendix B. One individual was a former teacher with curriculum development experience that now works in the main offices for the Board of Education. The other two applicants were both health and physical education teachers in the same middle school. One had prior experience in curriculum development, the other applicant had no prior experience but expressed motivation to gain more experience. This individual was experienced in computer and graphics programs that could be useful in the development of the

curriculum. After a formal twenty to thirty minute interview with each candidate, all three were hired in February 2006.

In March and April 2006, educational sessions with each curriculum writer were held at their respective schools to assist teachers with SCT and SCD facts. Specific topics that were covered in these sessions included information about the prevalence of the disease and trait, especially in the African American population. The genetics and inheritance pattern of the disease was covered, making a note of the 25% risk for two parents with SCT to have a child with SCD. The teachers learned about the specific shape of the red blood cells that are affected by gene mutations that give SCD its name, and why it causes many complications within the body. The complications and common characteristics of SCD were reviewed. It was made clear the difference between healthy individuals with SCT, and those affected by SCD. This helped relate back to the idea of inheritance, and why it is important for individuals to be aware of their trait status. Because they are healthy individuals, the only way to tell is through a simple blood test, and for many of the students in middle school, they were tested at birth and just need to contact the hospital to learn of their trait status. The different treatments for SCD, as well as the potential for a cure with bone marrow transplantation were explained. The curriculum writers were allowed to keep all informational materials including brochures, handouts, and Powerpoint presentations.

During the months of March and April, the curriculum writers took the information they learned about SCT and SCD and combined it with their knowledge of curriculum standards to create a curriculum appropriate for 6th, 7th, or 8th grade health classes. Once the first draft was finished, the summer was spent proofreading the curriculum for spelling errors and grammatical mistakes. When the teachers began working again in the fall, the project was able to resume.
Working together, the curriculum was corrected for factual errors and informational oversights. This allowed the teachers to learn of dependable resources, as well as of important facts to highlight when they are teaching their classes or presenting the curriculum to other teachers.

4.3 SPECIFIC AIM II: KNOWLEDGE OF SICKLE CELL DISEASE AND SICKLE CELL TRAIT

With a curriculum for the middle school level completed, the next step was to have health teachers use it in their classrooms. Instead of approaching teachers individually, it was decided that attending a teacher in-service day would be the best place to reach a large audience of health teachers at once. An in-service day is a day where there are no classes, and the teachers all gather together at one school for specialty training, and is a prime opportunity for new concepts or curriculum materials to be introduced. The group of teachers being targeted to teach the curriculum was health/physical education teachers for grades 6-8. Ideally there would be groups of teachers who work together at the same school that would all be willing to teach the new curriculum. CD-ROMs were distributed that included the entire curriculum for either computer based or paper-based teaching of the lesson. The teachers were also given a sheet that allowed them to evaluate the curriculum, and then return it to CHP. (Appendix C)

5.0 RESULTS AND DATA ANALYSIS

5. 1 SPECIFIC AIM I: PROVIDING ACCESS TO NEW EDUCATIONAL MATERIALS

The final curriculum for middle school health classes consists of three lessons. The curriculum can be distributed on a CD-ROM, and contains all the lessons and additional information in separate folders on the CD-ROM. There is a 'General Information' folder that contains an introductory statement for the teachers, as well as fact sheets for the teachers and the class about SCT and SCD. (Appendices D, E, and F) The folder for lesson one contains a lesson outline, a video about SCD, and a worksheet with an answer key to go along with the video (Appendices G and H). Lesson one includes reviewing some basic facts about sickle cell disease (with the use of the provided fact sheets), watching a 37 minute video, completing a worksheet that accompanies the video, and an assignment that involves writing a letter to an 'absent' classmate with 10 facts about SCD or SCT. The video is entitled, "Sickle Cell Disease: The Forgotten Disease" and was co-produced by the Dr. Spock Company and the Discovery Health Channel, and was originally aired on the Discovery Health Channel. The video follows a young girl with SCD and her pursuit for a cure for SCD. The video also has an interview with Tionne Watkins, a member of the singing group TLC, as well as an interview with a family whose daughter was successfully treated for SCD with a bone marrow transplant with her brother as a

donor. There is a worksheet that follows along with the movie, to help maintain the students' attentions.

Lessons two and three involve the students using online and other resources to make a poster that they will present to the class. The lesson two outline lists several online resources for the students to access (Appendix I). The third lesson/class period allows more time for research and creating the poster. Additionally, the inheritance pattern of SCD and SCT is emphasized, and a worksheet with answer key is included to help further teach the genetics of the disease. (Appendices J and K) A suggested grading rubric for the posters is enclosed if the teachers do not already have an established grading guideline. (Appendix L)

Also included on the CD-ROM is an extra credit folder that includes a worksheet (with answer key) that asks questions about the heterozygote advantage and malaria, as well as questions about why there are more people with SCT than SCD. This worksheet involves applying the knowledge they have already learned. (Appendix M)

5.2 SPECIFIC AIM II: KNOWLEDGE OF SICKLE CELL DISEASE AND SICKLE CELL TRAIT

In January of 2007, a presentation was made at a teacher in-service day. At this in-service day, the presentation on SCT and SCD curriculum was optional. Four teachers attended the SCD presentation, however, one was a fourth grade teacher with a personal interest, and the curriculum may not be at an appropriate learning level for her students. The other three teachers participated actively in the presentation, but said they would not have time to teach the curriculum, and could not take a CD-ROM or evaluation sheet with them.

In February of 2007, another presentation on the SCD curriculum was scheduled for an in-service day. This time the presentation was one of the main classes of the day, and was a required event. Over 20 teachers were in attendance, and many were interested in taking a copy of the CD-ROM with the curriculum. Seven teachers signed their names, email addresses, and school name on a sheet to be contacted in the future.

Attempts were mad to re-contact these seven volunteers, and four teachers responded (two teachers from South Brook Middle School, and two teachers from Carmalt Academy of Science and Technology). After looking over the curriculum, they were able to fill out the evaluation sheets (Appendix C) and return them. This evaluation assessed the amount of information provided, the appropriateness of the curriculum, the materials that were provided, the comfort level of the teacher, and the relevance of the curriculum. Each teacher also recorded what grade levels they teach. (See figure 6)



Figure 6: Grade levels being taught the SCD curriculum

The evaluation sheet consisted of eight questions, and the teachers circled the appropriate responses on a five point Likert Scale. Choices were 1 through 5, with an answer of 1 indicating 'not at all/strongly disagree', and an answer of 5 indicating 'definitely/strongly agree'. All teachers circled either 4s or 5s, with an answer of 4 indicating 'mostly/agree' about topics such as usability or satisfaction of the curriculum.

All teachers felt that there were ample materials provided to teach this curriculum (Question one, Figure 7). All teachers also reported that they felt this curriculum met the PA Academic Standards, and that the information is important for the students to learn (Questions 2 and 6, Figure 7). Teachers mostly felt the curriculum was appropriate for their grade levels, but one teacher reported that it may be too complicated for 6th grade students. The teacher added the lesson was still a good idea to introduce the topic of SCD and SCT (Question 3, Figure 7). The teachers also mostly felt that it was important to distinguish between SCT and SCD (Question 4,

Figure 7). As mentioned, all teachers felt this was important for students to learn, and most felt the information was relevant to their students (Question 7, Figure 7). One teacher noted that they felt it was mostly relevant to their students with an African American ancestry. The ethnic backgrounds of the two schools can be seen in Figure 8 and Figure 9, showing that while African American students make up a large portion of each school, they are not the majority. All teachers recorded that they mostly felt comfortable teaching the inheritance pattern of SCD to their students, and none of them felt 100% confident about teaching inheritance and genetics (Question 5, Figure 7). After reading through the entire curriculum, all teachers said they would definitely use this curriculum (Question 8, Figure 7).



Figure 7: Average Answers to Evaluation Questions

Table 1: Summary of questionnaire questions

Question 1	Satisfaction of material provided
Question 2	Curriculum meets academic standards
Question	
3	Material appropriate for the grade level
Question 4	Importance of emphasizing difference between SCT and SCD
Question 5	Comfort level teaching the genetics and inheritance of SCD
Question	
6	Importance of information
Question	
7	Relevance of information
Question	
8	Will they use the curriculum?



Students at Carmalt Academy of Science and Technology

Figure 8: Ethnicity of students at Carmalt Academy of Science and Technology



Students at South Brook Middle School

Figure 9: Ethnicity of students at South Brook Middle School

The second part to the evaluation sheet was more qualitative. The first question asked about the materials provided, and if any resources were needed. One teacher said they would give the CD-ROM to another teacher 'as is', and thought there were plenty of materials provided. Another teacher noted that creating a line at the top of each worksheet for the students' names would be helpful, and another teacher said they would like a VHS copy of the video because their school does not have the equipment to watch a video from a computer. Another teacher said that the initial summary of the lessons was an excellent addition. They also felt positive about the assignment of writing a letter to an 'absent classmate', and found it a creative exercise. All the teachers liked the worksheet accompanying the movie, it gives the students something to do and therefore holds their attention.

The second question asked why a teacher would or would *not* use this curriculum. Reasons stated why they would use the curriculum included the relevance to the students in their classes (specifically those with African American ancestry, noted as a "high risk" population by one teacher), and the possibility of having a child with SCD in their class (one teacher currently has a student with SCD). The main reason listed as to why they would *not* teach the curriculum was their lack of time. In middle schools, health and physical education are combined and only meet every third day. Two of those days are used for gym classes, and the third day is a health lesson. This means they are teaching health approximately twice a month. Associated with this issue, is the fact that they have to cover topics, such as drugs and alcohol, HIV/AIDS, nutrition, sex education, first aid, growth and development, and mental health (Answered by the teachers in question 3 of the qualitative questions). The teachers felt that when given the option to teach topics like those listed above, they would have to choose those because they are more general, and applicable to everyone. Teaching about SCD can be relevant to many people, but teaching about one specific disease instead of a general topic is less practical.

When asked what they would to do make the lessons more interesting for the students (question 4), one teacher suggested a quiz to go along with each lesson to keep the students on task. Another teacher suggested that instead of having them do research and present a poster, that they should write a research paper on the topic.

The final qualitative question asked about the teachers' personal experience with online curriculums. One teacher has written a curriculum on HIV/AIDS and shared it online, but was unaware if others had used it. The majority of the teachers said it may be a handy tool, but question the accuracy of the information presented. One teacher said they would absolutely never use a curriculum they found online.

6.0 **DISCUSSION**

6.1 SPECIFIC AIM I: PROVIDING ACCESS TO NEW EDUCATIONAL MATERIALS

This project has shown that when an organization such as the Pittsburgh Public School System is presented with an opportunity for specialized education, they are a receptive audience. There are two main reasons for this. The first being they are busy organization and the second is they are a growing organization. However, while it may be easy for the leaders of an organization to welcome a partnership and a new project, it is difficult to get a project of the ground for many reasons. It is not easy to ask an over-worked staff to do more, even though there is constantly new information and exciting approaches to teaching that are appearing. The schools system is willing to outsource some tasks to further increase and supplement existing curricula, as demonstrated by the hiring of Kaplan. Kaplan was not hired to create a new curriculum in health classes. Sometimes it takes an outsiders perspective to notice areas overlooked in a curriculum. The majority of the students in the Pittsburgh Public Schools are of African American ancestry; however, the majority of students do not know about SCD or SCT. The Pittsburgh School System was very receptive to the opportunity offered by CHP. When brought to their attention, it was agreed that SCD and SCT were topics of mutual interest.

The project also demonstrates that it is necessary to have one person from within each organization coordinating efforts. Sarah Martin proved essential in knowing and learning the procedures to get things accomplished through the school system. Appropriate paperwork and contacts within the department assisted with the project. The CHP group provided dependable resources and background knowledge of SCT and SCD. They ensured the curriculum contained all of the correct information. Contact was maintained through email, phone conversations, and meetings at the Board of Education's buildings or at the curriculum writers' school buildings.

This project was limited by the fact that only three teachers applied to help write the curriculum. However, all three people were motivated and qualified. Each writer contributed to the creation of this curriculum. Since the creation of the curriculum, one of the teachers has since switched schools, providing yet another site to implement the curriculum. It was challenging to contact teachers due to their complex schedules; however, when they were available to meet, they were very attentive and ready to participate in the project to make a curriculum that everyone would want to use.

Answer keys for the worksheets and fact sheets for the teachers were added as a convenience for teachers. SCD is a subject that not only do the students not know a lot about, but the teachers may have a limited background in the subject as well. The fact sheets were to help the teachers prepare for their classes. The answer keys were for them to check their students' work, and also to help them confirm their knowledge of this new topic.

6.2 SPECIFIC AIM II: KNOWLEDGE OF SICKLE CELL DISEASE AND SICKLE CELL TRAIT

With the limited (n=4) feedback from the teachers, it appears that the curriculum that was created by their peers was very well received. They all praised the final product, especially the ease of it all being organized and placed on a CD-ROM. The teachers that did not take a copy of the curriculum still complimented the final product, and several noted that they would have liked to use it, but lacked the time. Time is the most significant challenge for implementing the lesson plans. One teacher discussed the point that 9th graders have health class for a full semester, and it may be easier to introduce this new curriculum to those teachers. It is understandable that middle school teachers are forced to choose even among basic components of health, and adding in the topic of a single specific disease is something that is probably not high on their priority list.

Based on email receipt confirmations, it was known that all teachers (n=7) were receiving the emails that had been sent; however, only 4 responded. Of these four, only two had actually used the curriculum, while the other two had plans to use the curriculum, or its elements. No teacher used the entire curriculum presented to them, stating time as the major factor. The teachers that had used the curriculum reported that their students were interested, asked many questions, and even said they learned new information.

Teachers have obvious time challenges, having only 2 to 3 class periods per month for health instruction, and they are also lacking time to prepare lesson plans. The school district is under staffed and has a small pool of substitutes to hire on a day to day basis. Many times,

teachers would have to give up their "prep" period to fill in for an absent teacher. This limited their time to read over the curriculum, and the time to meet to discuss either the writing of the curriculum or the implementation of the curriculum. On one day, a teacher shared that out of a staff of 35 in their school, 7 teachers had called in sick, and only 2 substitute teachers were able to come to the school that day. This caused all the other teachers in the school to have to sacrifice their prep periods or to combine two classes to teach at once.

There was also a limited amount of teachers to participate in the evaluation of the curriculum. All of the feedback was positive, and many good suggestions were given about additions or corrections that could be made to the curriculum. Unfortunately, a sample size of 4 prohibits an accurate assessment of the materials.

It may have been more productive to directly approach teachers on an individual basis to check their availability and willingness to use the curriculum. Groups of teachers could be targeted by picking a school that has a high percentage of students with African American ancestry. Also, teaching by CHP staff may be considered. Guest speakers are always welcome in the classrooms.

It also appears that teaching a 9th or 11th grade class could be more effective. Students have health for an entire semester. In these grades it is not combined with physical education classes as in middle school. These classes would also have more time to complete all three lessons included in the curriculum, where as the middle school classes do not have time for these lessons.

Teachers appeared to be somewhat uncomfortable teaching genetic concepts to their students. While observing the lesson being taught in one class, the teacher became flustered and attempted to defer the teaching. In the Pittsburgh city schools, genetics is not taught until high

school (it is taught in 6th grade in some county schools). Perhaps more time should have been spent on creating exercises and work sheets about genetics and autosomal recessive inheritance to assist the teachers.

An online curriculum may be a viable option for classroom instruction. At the current moment there are no internet regulations regarding the quality of curriculum material. It is important to consider online forum lesson plans, due to the ease of application and potential strong interest from teachers.

6.3 CONCLUSIONS

This project showed the successful partnership of a growing public school system and an established pediatric hospital. The mutual collaboration provided the expertise in teaching and curriculum design coupled with factual information and details about SCT and SCD. CHP needed an outlet to share their information about SCT and SCD to make the community more aware of risk, especially information for teens as they approach reproductive age. This partnership with the schools has allowed CHP to reach a targeted age group at a vital time, and grants CHP the potential to continually reach this group.

This partnership also benefits the Pittsburgh Public Schools. The curriculum comes with a built in form of evaluation for the teachers to use while grading and judging the progress of their different classes. This curriculum is also sustainable. It was created by the teachers, and

provides the main facts about SCT and SCD. The program is easy to understand and use, and can be used for years to come without the need for CHP to come and guest lecture or observe. This allows the school system to have ownership of this curriculum. It is something of which they can be proud of because they helped create it, and can continue to perpetuate it for years to come.

In terms of the appropriate grade level for this curriculum, we found the school system does not allow an ample amount of time to teach this curriculum in a middle school. Based on time availability, and potential for learning more about genetics in a high school science class, this curriculum may be better suited for the high school level, specifically the 9th grade. Future directions, based on this information may include evaluating this curriculum with 9th graders, and adding more information and adjusting it for the 11th grade level of learning.

When approaching teachers, it may be better received by someone that only teaches health in a semester-long class (not health and physical education). Directly approaching teachers, instead of relying on them to attend informational meetings may increase use of the lesson plans. This provides for a personal meeting, but it also allows for a specific school or group of teachers to be chosen, perhaps a school that has a majority African American population.

This project has shown that SCD and SCT are topics that are important to teach to teens, and that teachers can be receptive to using a curriculum designed to educate these youth. This project also shows that the combination of two different institutions can produce a quality educational tool that can and will be used by public schools.

APPENDIX A

BOARD ACTION INFORMATION SHEET

Sarah Martin

Submitted By:

Action

Item #

General Authorization

December

2005

Action

Month

Lawrence

Hubbard

Person

Accountable

Action Requested:

Request for authorization to accept a donation of \$546.48 from Children's Hospital to engage three to four middle school teachers to develop supplemental materials relating to Sickle Cell trait and disease Between January 2006 and April 2006. Pittsburgh Public School Teachers will develop these materials to be used in the middle schools. Teachers will be compensated at the prevailing workshop rate of 22.71 an hour for up to six hours each. Children's hospital received this grant from the department of Health and Human Services to develop strategies to educate schools and communities about the importance of the awareness of Sickle cell disease and trait. These materials align with our Health, Safety and Physical Education standards.

Further Resolved:

That the Board expresses its appreciation and thanks to the Children's Hospital, and its sponsors for this generous gift to the district.

APPENDIX B

SAMPLE RESUME

I. <u>CERTIFICATION</u>	Pennsylvania Secondary Instructional II Certificate 1994	
	Areas of Certification:	General Science and Biology
	Emergency Certification	- 1998 to 2000
	Areas of Certification:	Physics, Chemistry
	Religious Certification -	Secondary - March, 1987

II. EXPERIENCE

Pittsburgh Public Schools	S Connelley <u>Administrative Center</u> 2000 -present	
	Secondary Science Resource Teacher	
Mount Lebanon Academy	Private tutor 1999 -p	oresent
	Biology, Physics, and General Science	
Pittsburgh Public Schools	Carrick High School1998 to	> 2000
	Biology, Physics, and General Science	
	Assistant Wrestling Coach	

Reizenstein Middle School		1992-1998
General Science		
PJAS-Sponsor	and	Judge
Head Wrestling Coach		

Diocese of Pittsburgh	Vincentian High School1991-1992
	Chemistry, Environmental Science, Algebra
	Staff Member - Middle States Evaluation
	Moderator - Environmental Club

III. PROFESSIONAL DEVELOPMENT

Research Experience for Teachers	Engineering and Product Innovation . 2005-6
	Design –based curriculum writing
Math Science Partnership	Science Curriculum Frameworks 2003
RMC Research Corporation	Professional Development Reviewer 2002-3
Pittsburgh Tissue Engineering	Teacher Education Program 2002
	Summer Intern
Administrative Summer Leadership	Secondary Science Facilitator 2001-2

PPS site coordinator and participant ... 2001

Teachers Teaching with Technology TI Graphing Calculator workshop... 2000-1

Yale-New Haven Teachers Institute Pittsburgh Planning Committee Member1998

National Demonstration Project

Presenter and participant

Harvard Graduate School of Education

Performance Assessment Summer Institute ... 1994

Outstanding Achievement in Science Education Award......1993-4

Middle School Teachers Science Training

Instructor-Cooperative Learning1993

IV. <u>EDUCATION</u> Duquesne University: Bachelors of Science, 1990

Major: Secondary Education - Science

Minor: Psychology

Content Area

General Biology I & II	Organic Chemistry
General Chemistry I & II	Recombinant DNA Technology
General Physics I & II	Genetics
Microbiology	Molecular Genetics
Microbial Ecology	Stress and Adaptation
Animal Development	Weather and Man

Masters Equivalency – Bachelors + 30 credits

APPENDIX C

EVALUATION FORM FOR TEACHERS

Teachers, please mark the number that you feel is most appropriate for the question being asked. 1= not at all, 2= a little, 3= unsure, 4= mostly, 5= definitely

1. I would feel comfortable teaching this unit with the materials that have been provided to me

1 2 3 4 5

 This curriculum helps meet the PA Academic standards ("Pennsylvania's public schools shall teach, challenge and support every student to realize his or her maximum potential and to acquire the knowledge and skills need to: Analyze how personal choice, disease and genetics can impact health maintenance and prevention." (10.1.9.E))

1 2 3 4 5

3. This material is appropriate for the grade level that I teach (your grade(s)_____)

1 2 3 4 5

4. It is important to emphasize the difference between Sickle Cell Trait and Sickle Cell Disease

1 2 3 4 5

5. I feel comfortable teaching genetics and the inheritance pattern of sickle cell disease

1 2 3 4 5

6. I think this information is important for my students to learn

1 2 3 4 5

7. I think is information is relevant to my students

1 2 3 4 5

- 8. I will use this curriculum
 - 1 2 3 4 5

1. What other materials would you like provided to you?

2. Any specific reasons why you would or would not use this curriculum?

3. What other topics do you cover in your health class?

4. How would you make this unit more interesting for your students?

5. Have you ever used curriculums that you found on line? Would you? If so, what did or didn't you like about them?

APPENDIX D

TEACHER INFORMATION



The Sickle Cell Disease Unit can be taught as part of the Communicable and Chronic Disease Unit. It is considered a chronic health condition. It can also be taught with the area of hereditary disease.

This Unit is set up for 2-3 lessons on the topic. Each lesson is designed with variations to permit adaptation to your own class/school needs. Students can work as a class or individually on a computer. All lessons can be online or printed out as needed.

All the information you as the teacher need is included in this packet. Please feel free to adjust as needed.

Standards

Pennsylvania Health Education

Standards 10.1 Concepts of Health-Health Problems and Disease Prevention

Standards 10.2 Healthful Living – Health Practices and Decision Making Skills

National Health Standards

Standard 1– Comprehend concepts related to health promotion and disease prevention.

Standard 3 – Demonstrate the ability to access valid health information.

Standard 5 – Demonstrate the ability to use decision-making skills to enhance health.

Overall Objectives

The student will be able to:

Explain and discuss what Sickle Cell Disease is, how it transmitted, symptoms and treatments.

Demonstrate their knowledge by designing and poster or game with pertinent information as instructed by the teacher.

Overview of Lessons

Lesson 1 – Informational Introduction, video, questions and answers

Lesson 2 – Informational Investigation and Research for poster or game

Lesson 3 – Design a Poster or Game for Active Participation

Teacher Resources

Informational Sheet # 1

Power point from Children's Hospital of Pittsburgh

Informational Sheet from Children's Hospital

Online Resources for Students

www.teenshealth.com Teens Health Education

- <u>www.sicklecelldisease.org</u> Sickle Cell Disease Assoc. of America
- o <u>http://www.state.nj.us/health/fhs/sicklecell/index.html</u> State of New Jersey
- <u>http://sickle.bwh.harvard.edu/menu_sickle.html</u> Harvard Univ.

Lessons Plans

Lesson 1: Introduction and Overview of Sickle Cell

Video and Questions

Lesson 2 : Web Search for Information to design poster

Lesson 3 : Completion of poster and Facts on Heredity

Resources

http://sickle.bwh.harvard.edu/menu_sickle.html Harvard education

http://www.scinfo.org/sicklept.htm Georgia Grady Health System

http://www.medicine.wustl.edu/~ysp/curriculum/gene 2b.htm Young scientist

program curriculum

http://www.stjude.org/phecom/0,2777,632 3505,00.html St. Jude Hospital

APPENDIX E

Facts For Teachers

Sickle cell disease is an **inherited blood disorder** affecting red blood cells. Normal red blood cells contain hemoglobin A. People with sickle cell disease have red blood cells containing mostly hemoglobin S, an abnormal type of hemoglobin. These red blood cells become sickle-shaped (crescent-shaped), and have difficulty passing through small blood vessels. There are several different types of **sickle cell disease**, the most common types are **homogzygous sickle cell disease** (SS disease), **sickle cell-hemoglobin C disease** (SC disease) and **sickle-cell beta thalassemia** (Sß⁺or Sß+⁰disease).

How do people get sickle cell disease?

- Sickle cell disease is not spread like a cold and cannot be caught from another person.
- It is an inherited condition.
- Sickle cell trait is a carrier condition for sickle cell disease .
- Sickle cell trait originated many years ago in areas of the world where malaria was present.
- People with **sickle cell trait** inherit one gene for normal hemoglobin A and one gene for defective hemoglobin S.

- People with **sickle cell disease** inherit a hemoglobin S gene from one parent and another abnormal hemoglobin from the other parent (i.e., Hemoglobin S, Hemoglobin C or Beta Thalassemia).
- If both parents have hemoglobin S *trait* there is a one in four chance with each pregnancy the child will have SS disease.
- If one parent has **hemoglobin S trait** and one parent has **hemoglobin C trait**, there is a one in four chance with each pregnancy the child will have SC disease.
- If one parent has hemoglobin S trait and one parent has beta thalassemia trait, there is a one in four chance with each pregnancy the child will have S⁶ or S⁰ disease.

Important facts about sickle cell disease

- Diagnosis of **sickle cell disease and trait** can only be determined by a special blood test.
- Comprehensive care includes early diagnosis, preventive measures, treatment of complications, and ongoing patient education.
- Many people with sickle cell disease live long and productive lives.
- Individuals with **sickle cell disease** can pursue a variety of vocations and professions.
- Many adolescents with **sickle cell disease** experience delayed puberty (the average delay is about two years).
- Yellowing of the eyes is common and should not be confused with hepatitis.
- The use of alcohol, "street" drugs and tobacco can greatly increase the risk of developing serious complications.

Sickle Cell Trait

- Approximately one in 12 African-Americans has sickle cell trait. The trait is also found in persons who come from Central America, Asia and the Mediterranean, but it can affect anyone regardless of ethnic background.
- Sickle cell trait affects the red blood cells.
- All red blood cells contain hemoglobin, which carries oxygen from the lungs to various parts of the body.
- People with sickle cell trait have both normal hemoglobin A and abnormal hemoglobin S in their red blood cells.
- People who do not have sickle cell trait or any other abnormal hemoglobin have red blood cells that contain only hemoglobin A.

- People with sickle cell disease have red blood cells that contain mostly hemoglobin S. Under certain conditions these red blood cells become sickle-shaped (crescent-shaped) and block circulation.
- Sickle cell trait rarely causes medical problems.
- People with sickle cell trait do not develop sickle cell disease.

Why is it important to know if I have sickle cell trait?

- Sickle cell trait in inherited from one's parents, like hair or eye color. If one parent has sickle cell trait there is a 50 percent (one in two) chance with each pregnancy of having a child with sickle cell trait.
- If both parents have sickle cell trait there is a 25 percent (one in four) chance with each pregnancy of having a child with sickle cell disease. Sickle cell disease is a lifelong illness that can result in serious health problems. For this reason, trait awareness is very important

APPENDIX F

Fact Sheet on Sickle Cell Disease

WHAT IS SICLE CELL DISEASE

- ✤ An inherited blood disorder that affects red blood cells
- Changed genes cause an abnormality of the hemoglobin
- The red blood cells lose their normal shape and appear sickle (half-moon) shaped (rigid and deformed)
- Misshapen (sickled) cells are not pliable and have difficulty squeezing through small blood vessels
- Abnormal hemoglobin changes the shape of the cell and causes it to lose its ability to carry oxygen all over the body

WHO IS PRONE TO SICKLE CELL DISEASE

- One in 400 hundred African-Americans is born with a form of a hereditary blood disorder
- ✤ In the U.S. sickle cell disease and trait are found primarily in people of African descent.
- Other individuals, especially from the Mediterranean, Middle East or India can also have this disorder, but it can affect anyone.

WHAT IS SICKLE CELL TRAIT

- ✤ A person is a carrier of the sickle cell trait, this means they have one working copy of a gene, and one non working copy of a gene
- Persons with the trait do not get ill
- ✤ 1 in 12 African Americans carry the trait

HOW DOES SICKLE CELL AFFECT PEOPLE

- Red blood cells cannot carry oxygen due to abnormal hemoglobin shape
- Red blood cells only survive 15 to 20 days in contrast with normal red blood cell that lives 120 days
- ✤ The sickle cell has difficulty getting through the small blood vessels
- The blood vessels become blocked depriving the cells of oxygen and causing tissue damage and extreme pain

STATISTICS

- Since September 1992 all newborns are screened for hemoglobin disorders
- ◆ In children with sickle cell the highest mortality rate occurs in the first five years.
- ✤ Mortality rate in the 5 to 20 year old group is low
- Most people with Sickle Cell disease usually live into their 40s

SIGNS AND SYMPTOMS

- Usually appear before the age of one
- Vary considerably with the individual-some can be severely affected other stay mildly affected
- Pain crises
- Pain most often affects bones and joints
- Generally last three to five days, but can last a week or more
- Some people with sickle cell have frequent, severe painful episodes that can result in hospitalization
- Others can have long pain free time
- Sometimes a child has a series of episodes and then be pain free for months or years.
- Infection and Fever
 - Higher occurrence of bacterial infection, primarily pneumonia, sepsis, and meningitis
 - Infection is the primary cause of death in young children
 - Repeated sickling in the spleen damages the spleen (The spleen loses its ability to clean blood)
 - Pulmonary Disease
 - About one third of patients will develop acute chest syndrome
 - Some will have frequent crises, while others will have no pulmonary problems
 - Symptoms include pain in chest, difficulty breathing, cough, fever, etc.
 - Chronic Kidney Disease
 - Blockage to the small vessels of the kidneys cause the kidneys to lose the ability to concentrate(hold) urine
 - Can result in dehydration
 - Treatment includes drinking large amounts of juice or water to stay hydrated
 - Stroke
- Blockage of the small blood vessels in the brain
- Prevention includes biannual imaging of the brain

TREATMENT

- People with Sickle cell take penicillin and folic acid every day to prevent infection and keep healthy red blood cells in the body
- Once a painful episode begins the emphasis is on pain management.
 - Most pain episodes are treated at home with pain medicine, relaxation and drinking lots of water or juice
 - No medications prevent the pain crisis or stop it
 - Aspirin should be avoided

APPENDIX G

LESSON PLAN ONE

(Begins on page 63)
Lesson 1 Sickle Cell Disease	Number of Classes 1 of 3	
Introduction,		
Facts about Sickle Cell		
Objectives	Standard	
Students will be able to (SWBAT):	Pa.State Standards for Health, Safety and Physical	
1. Complete a Sickle Cell worksheet and	Education	
answer the questions correctly.	Standards 10.1 Concepts of Health-Health	
2. In a letter to an absent classmate Describe	Problems and Disease Prevention	
and discuss 10 facts about Sickle Cell Disease	Standards 10.2 Healthful Living – Health Practices	
(transmission, signs and symptoms, and treatments).	and Decision Making Skills	
Procedures		
1. Introduce Sickle Cell – Give an overview- use yeach	ner printouts as your resource	
 Video on Sickle Cell : <u>The Forgotten Disease</u> Show to the class as a group or on individual computers Activity to do with the video - Worksheet to accompany the video 		
3. Letter to Absent Classmate with 10 significant facts about Sickle Cell		
4. Wrap up.		
Materials Computer and LCD Projector for group p	presentation of the Video/Power point or computer stations to	
view video/power point		
Teacher Printouts,		
Video The Forgotten Disease, or Power point from	Children's Hsopital	
Printed/On computer Worksheet to accompany the video		
Printed form/on computer Letter to Absent Classmate		
Vocabulary anemia, sickle cell trait, crises, heredity		
Evaluation Completion on the Letter to Absent Classmate with 10 facts.		
Printed worksheet that accompanied the video		
Resources United Streaming Video, Children's Hospital Information for teachers,		

APPENDIX H

MOVIE WORKSHEET AND ANSWER KEY

Sickle Cell

The Forgotten Disease

- 1. On what continent is sickle cell most common?
- 2. About how many babies are born each year in the US with sickle cell?
- 3. How many African Americans have the sickle cell trait in the United States?
- 4. What is sickle cell anemia?
- 5. What happens during a pain crisis?
- 6. Name the drug that is used to help reduce the severity of sickle cell pain crisis?
- 7. In the movie; who are the two girls having the experimental transplants?
- 8. Nina's transplant was very successful. What happened? Is she totally cured?
- 9. Define: engraftment
- 10. What is the difference between the two girl's transfusions?
- 11. Infinity's immune system had complications? What happened?
- 12. Who is the star who has sickle cell?
- 13. What other painful diseases could benefit from the sickle cell research?
- 14. In your own words do you think the research with the mice will be beneficial in the long run? Explain why or why not.

Answer Key for Movie Worksheet

- 1. Africa
- 2. About 1,000
- 3.2 million

4. A genetic disorder that destroys the ability of the red blood cells to carry oxygen throughout the body

5. The red blood cells become hard and pointed and can get stuck in small blood vessels, preventing oxygen from getting to organs causing damage to the surrounding organs, and severe pain

6. Hydroxyurea

7. Infinity and Nina

8. Yes, she is cured. It replaced her diseased bone marrow with healthy bone marrow.

9. When a surgery to place tissue (bone marrow) from a donor into a patient works, and the new cells become a working part of the patient's body systems

10. Nina's transplant uses bone marrow from her brother. Infinity's transplant uses blood cells from a baby's cord blood that is not related to her. This transplant does not work.

11. Her body did not accept the new bone marrow, and her immune system became very weakened because of it

12. T-Boz from TLC (Tianne Watkins)

13. Leukemia

14. Answers will vary.

APPENDIX I

LESSON PLAN TWO

Lesson Sickle Cell Disease	Number of Classes 2 of 3	
This lesson will take two classes periods to		
complete		
1		
Objectives	Standard	
Students will be able to (SWBAT):	PA.State Standards for Health, Safety and Physical	
Design a poster to inform and educate others about	Education	
Sickle Cell Disease including transmission, symptoms,	Standards 10.1 Concepts of Health-Health	
and treatments	Problems and Disease Prevention	
	Standards 10.2 Healthful Living – Health Practices	
	and Decision Making Skills	
Procedures		
1. Brief review of Day 1 Check for Understandi	ng Refer to Teacher Printout for Questions	
2. Then on line article from Teens Health		
http://kidshealth.org/teen/diseases_conditions	/blood/sickle_cell_anemia.html	
www.sicklecelldisease.org Sickle Cell Dis	sease Assoc. of America	
http://www.state.nj.us/health/fhs/sicklecell/ind	lex.html State of New Jersey	
http://sickle.bwh.harvard.edu/menu_sickle.html Harvard Univ.		
3. Design poster with information		
Materials online resources sickle cell disease, paper, printer, poster paper, markers		
Vocabulary hemoglobin, sickle cell trait, crises, sickle cell disease		
Evaluation Completion on Sickle Cell Poster		
Resources online resources		

APPENDIX J

Lesson Sickle Cell Disease	Number of Classes3 of 3			
Complete Sickle Cell Poster				
Consting and Sigkle Coll				
Genetics and Sickle Cell				
Objectives	Standard			
Students will be able to (SWBAT):	Pa. State Standards for Health, Safety and Physical			
1. Design a poster to inform and educate others about Sickle Cell Disease including, transmission	Education			
symptoms, and treatments	Standards 10.1 Concepts of Health-Health			
2. Explain now Sickle Trait is transmitted	Problems and Disease Prevention			
	Standards 10.2 Healthful Living – Health Practices			
	and Decision Making Skills			
Procedures				
1. Brief review of Day 2 Check for Understanding Refer to Teacher Printout for Questions				
2. Using the Handout on genetics explain how sickle cell trait is transmitted. You can make an overhead of				
Genetic handout or you can have students do the worksheet				
Materials Genetic handout,				
Computers for web search, Books and Magazine, Poster Paper, Markers, Glue, Scissors				
Students can use Microsoft Publisher to complete the	neir poster			
Vocabulary Sickle Cell Disease, Sickle Cell trait, crises, hemoglobin				
Evaluation Completed posters will be displayed in classrooms and around school				
Resources Web Quest				

LESSON PLAN THREE

APPENDIX K

Genetics and Sickle Cell

Probability of Getting Sickle Cell Disease

Case One

Both parents are carriers of the sickle cell trait. They each have one normal copy of the gene (the heart), and one abnormal copy (triangle). What does this mean for their children?

Parent 1

Δ ¥

Δ ¥

Parent 2

Kid

If the kid receives either the heart (a normal gene) or the triangle (the sickle cell gene) from each parent, how many combinations are possible? Draw them below, and label whether the kid is unaffected, a carrier, or has sickle cell disease in each drawing.

Case Two

One parent has sickle cell disease and the other is unaffected. What does this mean for their children?

Parent 1 Parent 2

 $\checkmark \checkmark \qquad \Delta \quad \Delta$

Kid

If the kid gets one of the two possibilities from each parent, how many combinations are possible? Draw them.

What conclusions do you get?

Answer Key for Genetics Sheet

Case one:

4 possible combinations

Two hearts (healthy)

Hearth/triangle (trait)

Triangle/heart (trait)

Two triangles (disease)

Case Two:

Hearth/triangle (x2)

This means that each child will have the trait, it will not be possible for them to have sickle cell disease, or have two healthy genes. Every child that these parents have together will have sickle cell trait.

APPENDIX L

Making A Poster : Disease Poster Rubric

CATEGORY	4	3	2	1
Title	Title can be read from 6 ft. away and is quite creative.	Title can be read from 6 ft. away and describes content well.	Title can be read from 4 ft. away and describes the content well.	The title is too small and/or does not describe the content of the poster well.
Required Elements	The poster includes all required elements (symptoms, cause, transmisson, and treatments) as well as additional information.	All required elements are included on the poster (symptoms, cause, transmisson, and treatments).	All but 1 of the required elements are included on the poster.	Several required elements were missing.
Effectiveness of Graphics	All graphics are related to the disease you researched and make it easier for others to understand.	All graphics are related to the disease you researched and make it somewhat easier for others to understand.	All graphics relate to the disease you researched.	Graphics do not relate to the disease you researched.
Knowledge Gained	Student can accurately answer all questions related to facts in the poster and processes used to create the poster.	Student can accurately answer most questions related to facts in the poster and processes used to create the poster.	Student can accurately answer about 75% of questions related to facts in the poster and processes used to create the poster.	Student appears to have insufficient knowledge about the facts or processes used in the poster.
Quality of Poster	The poster is exceptionally attractive in terms of design, layout, and neatness.	The poster is attractive in terms of design, layout and neatness.	The poster is acceptably attractive though it may be a bit messy.	The poster is distractingly messy or very poorly designed. It is not attractive.
Grammar and Mechanics	There are no grammatical errors on the poster and it has been edited for spelling and punctuation.	There is 1 grammatical or mechanical error on the poster.	There are 2 grammatical or mechanical errors on the poster.	There are more than 2 grammatical or mechanical errors on the poster.

APPENDIX M

EXTRA CREDIT AND ANSWER KEY

SICKLE CELL ASSESSMENT—Extra Credit

Name_	Date	

Class_____ Homeroom _____

1) Label the red blood cell you think is normal and which would be found in someone with sickle cell disease.



2) Why is sickle cell disease a problem? (What happens that shouldn't when someone has the disease?)

3) How is sickle cell disease associated with malaria?

4) Check the line if the person suffers from the painful symptoms of sickle cell anemia.

____ The person has sickle cell disease.

_____ The person is a carrier for sickle cell.

- _ The person doesn't have sickle cell and doesn't carry the trait either.
 - 5) Explain why so many more people are carriers than actually have sickle cell disease and the effects this can have on others.

Extra Credit Answer Key

- 1. Cell on left if normal, cell on right is sickle
- 2. Red blood cells lose the ability to carry oxygen. This causes them to change from a round, soft shape, to a hard and sickled shape. These sickle cells can get stuck in small blood vessels, blocking the vessels and preventing any oxygen from getting through to the organs. This causes extreme pain, and even organ damage.
- 3. Someone with sickle cell trait is more resistant to malaria. This is why sickle cell disease is more common in areas where malaria was a problem, such as Africa, the Middle East, and India.
- 4. The person has sickle cell disease
- 5. So many more people are carriers than actually have disease because it takes two carrier parents to have a 25% chance to have a child with disease. It only takes one parent with sickle cell trait (a carrier) to have a 50% chance to have a baby with sickle cell trait.

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