A Statewide Needs Assessment:

On the Path to Developing Tailored Sickle Cell Trait Education in the State of Nevada

Yasmine A. Mohamednur, MPH Candidate

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Trudy Larson, MD

Advisor, Graduate Committee Chair

Mary Pennington, BA

Field Experience Supervisor, Graduate Committee Member

Echezona Ezeanolue, MD, MPH

Graduate School Representative, Graduate Committee Member

Elizabeth Christiansen, PhD

Graduate Committee Member

Assembly Committee: Health and Human Services
Exhibit: R Page 1 of 79 Date: 03/29/2019
Submitted by: Yasmine A. Mohamednur

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Abstract

Background: Sickle cell trait (SCT) is an inherited condition that affects the hemoglobin in the red blood cells, and it is usually considered clinically mild. However, adverse health outcomes have been associated with some individuals who have SCT, particulary in low oxygen situations and with overexertion. Due to the generally benign nature of SCT, individuals with SCT and their families typically receive minimal education on the potential health risks that are associated with SCT.

Objectives: (1) Determine existing knowledge and awareness of potential health risks associated with SCT among parents of children with SCT and medical providers in the State of Nevada. (2) Determine preferred educational approaches of parents and medical providers to develop tailored SCT education.

Methods: Using the PRECEDE-PROCEED model as a general context, a cross-sectional survey design was used to obtain information from parents of children with SCT and medical providers on SCT education needs in the State of Nevada. Two key informant interviews were also conducted. A total of 600 surveys were sent to providers and parents statewide. Quantitative analyses were performed on all close-ended survey questions. Qualitative thematic analyses were performed on open-ended survey questions and key informant interviews.

Results: One hundred and twenty-one surveys were returned and eligible for analyses. Many parents surveyed did not believe that they had adequate support services or educational information for their child's SCT. Medical providers had an adequate understanding of SCT, but a majority of responders indicated that they never or rarely saw patients with SCT. In terms of educational delivery needs, both parent and provider respondents preferred an educational delivery method that was online, such as a website with dedicated resources to SCT.

Conclusions: There is a significant need for SCT education and resources in the State of Nevada. Provision of educational information can be accomplished in several ways, and this study has provided insight into the preferred methods of educational delivery and topics of interest from a sample of individuals who are highly affected by SCT within the State of Nevada.

Background

Sickle cell trait (SCT) is an inherited condition that affects the hemoglobin in the red blood cells, and it is usually considered to be clinically mild. Hemoglobin is the iron-containing protein that transports oxygen in the red blood cells to the rest of the body. SCT is characterized as being heterozygous for the sickle cell gene (HbS), meaning an individual inherits a normal hemoglobin gene (HbA) from one parent, and an abnormal or mutated hemoglobin gene (HbS) from the other parent (Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009). SCT is different from sickle cell disease (SCD), and to understand SCT it is essential to realize the difference between the two. SCD is a group of inherited red blood cell conditions (including HbSS, also known as sickle cell anemia), where an individual inherits two abnormal hemoglobin genes (Bloom, 1995). In the instance of sickle cell anemia, individuals are homozygous for the sickle cell gene, meaning they inherit one sickle gene from each parent. This mutation in hemoglobin genes results in an abnormal hemoglobin protein which leads to the distortion of the red blood cell from a round shape to a crescent or "sickle shape" (Bloom, 1995).

The sickle shape of the red blood cell associated with SCD was first described in Western medical literature in the November 1910 edition of *The Archives of Internal Medicine* in an article titled, "Peculiar Elongated and Sickle-Shaped Red Blood Corpuscles in a Case of Severe Anemia" (Savitt, 2010). The article was written by Cardiologist James B. Herrick and his intern Ernest E. Irons after they reviewed the blood cells of a twenty-year old dental student from the island country of Grenada who presented as weak, dizzy, short of breath, and had a painful ankle sore (Savitt, 2010; Washington, 2006). Herrick and Irons noted the shape of his red blood cells as thin, elongated, and sickled (Washington, 2006). In February of 1911 an article published in the *Virginia Semi-Monthly* cited Herrick's article and described a twenty-five year old African-American woman with similar symptoms as the dental student from Grenada; subsequently there were many other case reports fitting similar clinical symptoms associated with sickle-shaped red blood cells (Savitt 2010; Washington, 2006). It is now known that these

abnormally shaped red blood cells break down causing anemia and obstruction of blood vessels, leading to recurring episodes of severe pain and multiple organ ischemic damage (Creary, Williamson, & Kulikarni, 2007). In contrast to sickle cell disease, people with sickle cell trait are generally healthy (Creary, Williamson, & Kulikarni, 2007). Although sickle cell trait is considered to be clinically benign in most cases, adverse health outcomes have been associated with individuals who have sickle cell trait; particularly in low oxygen situations and heavy exertion activities (Centers for Disease Control and Prevention, 2013; Koopmans, Cox, Benjamin, Wright-Clayton, & Friedman-Ross, 2011; & Mitchell, 2007).

Sickle Cell Trait Inheritance

SCT is a carrier state, meaning that the individual can never develop sickle cell disease, but they risk having children with either SCT (HbAS) or SCD (HbSS). For example, children born to two parents with SCT have a twenty-five percent chance of having sickle cell disease (HbSS), a twenty-five percent chance of not being affected (HbAA), and a fifty-percent chance of having sickle cell trait (HbAS) (Bloom, 1995; Treadwell, McClough, & Vichinsky, 2006). If one parent has SCT and the other parent is not affected, half of the births will result in the HbAS gene, and half will result in the HbAA gene (Bloom, 1995). Given the examples of the SCT inheritance pattern, awareness of one's carrier status is important because SCT carriers can pass on their variant gene to their children. It is essential that individuals with sickle cell trait understand these implications.

Sickle Cell Trait Prevalence and Incidence

Approximately three hundred million people worldwide are SCT carriers, and it is estimated that four million people in the United States are trait carriers (HRSA, 2010). It is difficult to know the precise numbers of SCT carriers because many individuals do not know their carrier status (Grant et al., 2011). SCT is predominant amongst individuals with African ancestry. The prevalence of sickle cell trait is approximately one in every twelve births, or eight to ten percent, amongst African-Americans, and the prevalence is as high as twenty-five to thirty percent in certain areas of Western Africa (Vichinsky, 2012).

Although the trait is predominant amongst Africans and African-Americans, it is a common misconception that sickle cell trait and sickle cell disease only affect individuals of African ancestry. An individual of any ethnic or racial origin can have sickle cell trait, and SCT has been documented in individuals with ancestors from the Middle East, the Mediterranean, Eastern India, the Caribbean, and in Latino populations (Creary, Williamson, & Kulkarni, 2007). For example, at least 1 in 180 Hispanic children born in the United States have sickle cell trait, and 1 in 625 Caucasian children born in the United States are sickle cell trait carriers (Vichinsky, 2012).

Sickle Cell Trait Health Implications

Extensive literature points to the morbidity and potentially fatal outcomes that can be associated with sickle cell trait (Harris, Haas, Eichner, & Maron, 2012; Hédreville et al., 2008; Johnson, 1982; Key & Derebail, 2010; Rosenheim, 1970; Tiernan, 1999; Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009; Vichinsky, 2012). Considered rare, adverse health outcomes that have a definite association with sickle cell trait include: renal medullary cancer, traumatic hyphema (blood in the anterior chamber of the eye), hematuria (blood in the urine) , renal papillary necrosis (death of the renal papilla), loss of the concentrating ability of the kidney, splenic infarction (occlusion of splenic vascular supply), exertional rhabdomyolysis (muscle breakdown from exertion), and exercise-related sudden death (particularly in military recruits and young athletes) (Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009).

Research suggests that the pathophysiology underlying the complications of SCT are not known, but is probably related to the sickling of red blood cells with intense activity (Key & Derebail, 2010). Many of these adverse health outcomes can be brought on by dehydration, increased atmospheric pressure, low oxygen levels in the air, or high altitudes (Centers for Disease Control and Prevention, 2013). In many of these situations low oxygen is a key factor. Low levels of oxygen leads to a process called polymerization, where molecules aggregate on the red blood cell membrane and eventually lead

to hemolysis (red blood cell breakdown) and vascular occlusion (blockage of a blood vessel); which can lead to tissue ischemia and organ complications (Mitchell, 2007). During exertional activities, such as intense exercise, the serum pH decreases and the body temperature increases to allow for increased delivery of oxygen to the body (Mitchell, 2007). These internal changes lead to higher concentrations of deoxygenated hemoglobin (Mitchell, 2007). Individuals with sickle cell trait may not be able to compensate for these changes as would a person without the trait in a similar situation of overexertion. As Mitchell (2007) states, "Under usual circumstances, cardiac output is likely to increase substantially and raise perfusion pressure sufficiently to overcome local tissue hypoxia and avoid the membrane polymerization that can occur with sickle cell trait" (p. 300). Polymerization leads to a surge of events, including "…rhabdomyolysis, myoglobinuria, acute renal failure, release of vasoactive substances, disseminated intravascular coagulation and coronary vasoconstriction" (Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009, p. 510). For people with sickle cell trait, membrane polymerization related to vigorous activity and low oxygen levels may be the key in understanding adverse outcomes (Mitchell, 2007).

Perhaps one of the most well-known definite clinical associations with sickle cell trait is the potential for renal complications. In 1974 Dr. Leonard Berman described the six nephropathies (renal diseases) of sickle cell trait and sickle cell disease. As Berman stated in his article, "It is fitting that the shape of a sickled red blood cell (RBC) should resemble that of a kidney. The patient with sickle cell trait or disease is liable to experience several types of sickle cell nephropathy" (1974, p. 1279). These nephropathies, as described by Berman, include: "gross hematuria, papillary necrosis, nephrotic syndrome, renal infarction, hyposthenuria and pyelonephritis" (as cited in Baig, Lin, Rasheed, & Mittman, 2006, p. 1171). In the mid-1990s researchers added the seventh nephropathy, renal medullary carcinoma. This type of cancer is described as being "…a rare but extremely aggressive tumor that originates from the renal collecting ducts of young patients with sickle cell trait" (Baig, Lin, Rasheed, &

Mittman, 2006, p.1171). Renal medullary cancer is almost always metastatic at the time of diagnosis, and is usually unresponsive to therapy; the median survival is four months (Baig, Lin, Rasheed, & Mittman, 2006). Yet, what is most interesting about renal medullary carcinoma is the fact that almost all patients described with this type of cancer have had sickle cell trait, and have ranged in age from 10 to 40 years (Baig, Lin, Rasheed, & Mittman, 2006; Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009).

In addition to renal complications, recent literature indicates that sickle cell trait may be a risk factor for venous thromboembolism (VTE). A 2007 case-control study by Austin et al., indicated African-Americans with SCT had approximately a four-times increased risk for pulmonary embolism (odds ratio 3.9; 95% CI, 2.2-6.9) and approximately a two-times increased risk of both deep vein thrombosis or pulmonary embolism (odds ratio 1.8; 95% CI, 1.2-2.9) (as cited in Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009). Knowledge of these associations is essential for appropriate precautions for individuals with sickle cell trait.

Sickle Cell Trait and Athletics

The issue of adverse health outcomes associated with sickle cell trait has gained increasing attention in the athletic world, especially in college sports. As of April 2010 the National Collegiate Athletic Association (NCAA) has adopted recommendations for the universal screening of sickle cell trait for all Division I college athletes (Koopmans, Cox, Benjamin, Wright-Clayton, & Friedman- Ross, 2011). This was in response to the 2006 death of Dale Lloyd, a football player at Rice University who had sickle cell trait and died shortly after an intense football practice (Koopmans, Cox, Benjamin, Wright-Clayton, & Friedman- Ross, 2011). In August of 2012 the NCAA implemented universal screening for all Division II athletes, and there is a current proposal to implement universal sickle cell trait screening for all Division III athletes, for new student athletes in 2013, and all student athletes beginning in 2014 (NCAA, 2013). The morbidity related to sickle cell trait in professional athletic sports has also been reported. A classic

example was in 2007 when Ryan Clark, an NFL player for the Denver Broncos and sickle cell trait carrier, suffered a splenic infarction due to overexertion at high altitude; the field where he played was 5,280 feet in elevation (Jordan, 2012). As a result, Clark had to have his spleen and gallbladder removed (Jordan, 2012). Clark's story is one of many examples of athletes who have suffered health implications related to sickle cell trait and overexertion.

The Rice University player, Dale Lloyd, died of exertional rhabdomylisis (ER), where skeletal muscle degrades as a result of intense exercise (Koopmans, Cox, Benjamin, Wright-Clayton, & Friedman-Ross, 2011). Lloyd's death was not the first exercise-related death associated with sickle cell trait. In the United States, there had been at least twenty reported football deaths in athletes with sickle cell trait between 1974 and 2010 (Koopmans, Cox, Benjamin, Wright-Clayton, & Friedman-Ross, 2011). In addition to exertional rhabdomylsis, SCT has also been linked to fatal exertional heat illness (EHI) after strenuous exercise. The main difference between ER and EHI is that EHI is related to heat and ER is not (Koopmans, Cox, Benjamin, Wright-Clayton, & Friedman-Ross, 2011). Yet, it is important to note that ER and EHI can occur in athletes with or without SCT.

The association of SCT and fatal EHI was first described in the early 1970's from an issue that had stemmed from military basic training. In 1969, four recruits for the United States Armed Forces died suddenly; all four of the recruits were African-American, and the autopsy results revealed that the men's red blood cells had been sickle shaped (Washington, 2006). The altitude of the basic training locations where the men trained were 4,060 feet in elevation (Washington, 2006). This suggested that the deaths might have been related to sickle cell crises, which were probably triggered by the low oxygen environment of the high altitude (Washington, 2006). The first studies to examine sickle cell trait and sudden death following the 1969 incident were conducted by Jones & Donowho (1970) and Rosenheim (1970). As a result of these studies some branches of the military began to exclude individuals with SCT from joining the military (Washington, 2006). These restrictions were removed in 1985, however there

were major ethical concerns regarding targeted screening and discrimination; in fact the issue of discrimination related to sickle cell trait status continues to be a major ethical concern related to sickle cell trait screening in present day (Washington, 2006). A larger follow-up study in 1987 by Kark, Posey, Schumacher, & Ruehle found "… approximately a 28-fold increased risk of exercise-related deaths (relative risk =27.6; 95% CI,9-100; P < .001), unexplained by preexisting illness among US Army recruits with sickle cell trait compared with those without sickle cell trait in their cohort study of 2 million enlisted military recruits" (Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009, p. 510). Rather than excluding individuals with SCT from military service Kark et al. recommended a universal approach. Kark et al. suggested that the same precautions should be taken for all military personnel to decrease the risk of exertional heat illness (EHI) because all military personnel were at risk for EHI, it was just that individuals with SCT were at increased risk for fatal exertional heat illness (Jordan, 2012).

Despite the noted associations in the medical literature between adverse health outcomes and sickle cell trait, there is no consensus among medical professionals as to whether sickle cell trait is really clinically mild or if it is a considerable health risk. The reasons for this debate are complex, many stemming from the potential ethical issues that surround labeling sickle cell trait as an illness. Regardless of these issues, there is universal agreement that sickle cell trait poses a significant public health risk because it affects millions of people worldwide, and there are potential adverse complications associated with the trait. As stated by Grant et al., (2011), "Although the issue of whether SCT is clinically benign or a significant health concern has not yet been resolved, the potential health risk to affected individuals is of vital importance and represents a tremendous challenge in protecting, promoting, and improving the health of approximately 300 million people worldwide and 3 million people in the US who possess the trait" (p. S435). One of the major public health implications related to sickle cell trait is status awareness, which can be accomplished through screening.

Sickle Cell Trait Screening

On May 16, 1972 President Richard Nixon signed the National Sickle Cell Anemia Control Act. The overall goal of the act was to bring more attention to sickle cell anemia by increasing funding for research and creating Comprehensive Sickle Cell Centers (National Heart, Lung, and Blood Institute, 2002). At the time "…SCD received less public and professional support compared with other less prevalent diseases, such as cystic fibrosis, despite having a substantial public health impact" (Smith, Oyeku, Homer, & Zuckerman, 2006, p. 1764). As a result of the Sickle Cell Anemia Control Act, seventeen states enacted sickle cell screening laws, and at the time, screening for sickle cell disease was a test usually done by employers (Washington, 2006). This attempt by the federal government to bring attention to sickle cell disease proved unsuccessful because the law also mandated sickle cell screening exclusively for the African-American population; and in doing so the act reinforced the flawed notion that sickle cell disease is solely a "Black disorder" (Washington, 2006). President Nixon described sickle cell disease in his statement regarding the National Sickle Cell Anemia Control Act as follows, "This disease is especially pernicious because it strikes only Blacks and no one else" (The American Presidency Project, 2013, para. 3). Additionally, the early screening programs created confusion by mixing sickle cell disease with sickle cell trait, and individuals with sickle cell trait were told that they had the disease (Washington, 2006). Individuals who were targeted by screening efforts questioned discrimination, and there was a critical lack of follow-up and education for individuals who were identified with either trait or disease at the time. These programs proved to be ineffective and were eventually terminated (Washington, 2006).

Today in the United States sickle cell disorders are screened for at birth through Newborn Screening Programs. Newborn Screening (NBS) is defined as a "Public health system comprised of screening, diagnosis, management, evaluation, and education" (Nevada Newborn Screening Practitioner's Manual, 2012, p.3). It is currently recommended by the American College of Medical

Genetics that State NBS programs screen for a panel of thirty-one core disorders and twenty-six secondary disorders as approved by the Discretionary Advisory Committee on Heritable Disorders in Newborns and Children (American College of Medical Genetics, 2006). The first statewide Newborn Screening Program for sickle cell disease was initiated in New York in 1975 (Benson & Therrell, 2010). In 1996, the American Academy of Pediatrics recommended that all 50 states implement targeted newborn screening for hemoglobinopathies (Long, Thomas, Grubs, Gettig, &Krishnamurti, 2011). In the same year, 1996, the US preventive taskforce (USPTF) reviewed the evidence for screening for sickle cell disease in newborns and recommended universal screening (US Preventive Task Force, 2007). Hemoglobinopathies refers to "A group of genetic conditions that result in the production of abnormal hemoglobin(s) or the lack of production of normal hemoglobin" (Benson & Therell, 2010, p.134). Both sickle cell disease and sickle cell trait fit in the category of hemoglobinopathy. Screening for SCT has finally become universal in the United States through newborn screening. Even though national recommendations have been in place since 1987, universal newborn screening for sickle cell disease and sickle cell trait has only been required in all fifty states, as well as the District of Columbia, since May 1, 2006 (Benson & Therrell, 2010). Prior to 2006, only some states screened for SCT, and as a result there are millions of Americans who are unaware of their SCT status (Centers for Disease Control and Prevention, 2013). The National Healthy People 2020 goals dedicated to SCT screening are centered on status awareness and education related to SCT; one of the goals states, "Increase the proportion of hemoglobinopathy carriers who know their own carrier status" (Department of Health and Human Services, 2013, BDBS-10).

Due to the potential reproductive and health implications of sickle cell trait, it is essential that individuals are aware of their SCT status. People who are encouraged to be screened for sickle cell trait outside of the established screening systems include: all expectant couples and those planning to have children, people with a family history of sickle cell disease, people with a family history of thalassemia,

and people with a family history of sickle cell trait (Jordan, 2012). Due to the generally benign nature of sickle cell trait and lack of medical consensus on the clinical significance of the trait, individuals with SCT tend to receive minimal follow-up services and education on the potential risks that are associated with sickle cell trait once they receive a positive screen. Additionally, some newborn screening programs provide parents educational materials related to sickle cell trait when a child is born, but the information related to potential health implications is inconsistent due to a lack of medical consensus regarding sickle cell trait severity. Given the potential array of adverse health outcomes associated with sickle cell trait, individuals must be aware of their SCT status and must be properly educated about the potential health risks as well as precautions to take in order to prevent adverse health outcomes.

Sickle Cell Trait Knowledge and Awareness

Few studies or surveys have examined the awareness of sickle cell trait or sickle cell disease amongst individuals or large communities. Prior studies that have examined strategies to enhance awareness of sickle cell trait or disease have focused on particular groups or issues such as African-American communities, or topics such as genetic counseling. For example, a 2005 cross-sectional telephone survey of African-American women ages 18-30 years old in St. Louis, Missouri assessed existing community knowledge regarding sickle cell disease utilizing the PRECEDE-PROCEED framework (Boyd, Watkins, Price, Fleming, & DeBaun). The results yielded strong evidence that African-American women were poorly informed about topics such as genetics related to sickle cell disease. In terms of educational preferences to learn about sickle cell disease many of the participants in this study favored pamphlets, educational meetings, radio, and television announcements. A 2011 study by Long, Thomas, Grubs, Gettig, & Krishnamurti examined the attitudes and beliefs towards genetic testing, prenatal testing, and newborn screening. The study also included an assessment of attitudes and beliefs regarding sickle cell disease, and any barriers to education and awareness. The study participants were African-American men and women whom the researchers recruited from flyers and an existing research

database in Pittsburgh, PA. The researchers noted that they did not investigate if the participants knew the difference between sickle cell disease and sickle cell trait. The study results indicated that many of the individuals did not know their own sickle cell trait status, and many had low perceived susceptibility to have a child with sickle cell disease. Furthermore, participants identified many personal and societal barriers to accessing sickle cell disease education. The study results also identified that active learning and community-based events were the preferred method of educational learning for these participants.

While there has been minimal research dedicated to investigating community level sickle cell disease and trait awareness, there are some community-based sickle cell awareness programs. A noteworthy program through St. Jude Children's Research Hospital provides community outreach education related to sickle cell disease and trait called KYSS (Know Your Sickle Cell Status). KYSS is "A community, faith-based program that uses teen-peer education to raise awareness among African-American teens at risk for having a child with sickle cell disease" (St. Jude Research Hospital, 2014, para. 3). Another program through St. Jude titled, STARR (Success through Academics, Research and Resources), "…helps public school teachers meet the special needs of students with sickle cell disease" (St. Jude Research Hospital, 2014, para. 4). Similar community outreach educational programs are available at other national comprehensive sickle cell centers that focus on sickle cell disease research, training, and education (National Institutes of Health, 2008). In 2012, the University of South Florida implemented a sickle cell trait educational initiative in collaboration with the National Athletic Trainers' Association (NATA) Ethnic Diversity Advisory Committee and the NCAA. The university created a website with educational resources aimed "…to educate coaches, healthcare professionals, parents, and student athletes on sickle cell trait and the prevention of sudden death [related to sickle cell trait]" (University of Southern Florida, 2012, para.1). Although there have been some educational initiatives related to sickle cell trait, there is still little information as to the effectiveness of these programs to raise awareness on the potential health implications related to sickle cell trait.

Sickle Cell Trait and Newborn Screening in Nevada

Nevada is a state with a rapidly increasing and diversifying population, and the number of individuals with sickle cell trait and sickle cell disease is steadily on the rise (Figure 1). In Nevada, newborn screening for hemoglobinopathies became required and available on July 1, 1990 (Benson & Therrell, 2010). Currently, screening for sickle cell disease and trait is done for all infants in the State of Nevada. On average, Nevada has approximately 34,000 births per year, and during the years 2005 through 2012, 5,326 Nevada newborns were identified as variant hemoglobin trait carriers (Nevada Division of Public and Behavioral Health Office of Public Health Informatics and Epidemiology, 2013). Of the 5,326 variant trait newborns, 2,957 had HbAS identified, meaning that these newborns were identified as sickle cell trait carriers.

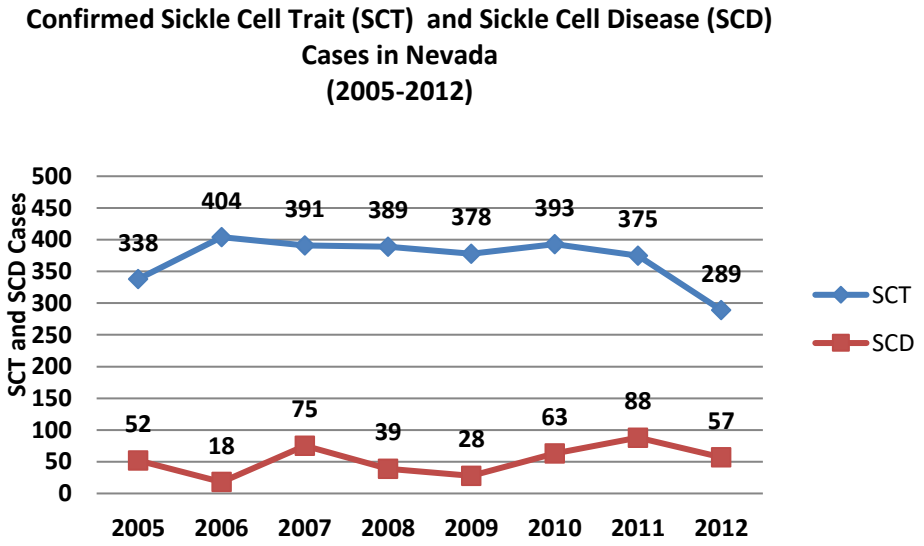


Figure 1: Confirmed SCT and SCD Cases in Nevada 2005-2012
Source: Nevada Division of Public and Behavioral Health, Newborn Screening Program (2013)

Project Overview

The Nevada Newborn Screening Program is under the Bureau of Child, Family, Community Wellness, Women's and Children's Health at the Nevada Division of Public and Behavioral Health. The purpose of the Nevada Newborn Screening Program is to, "…decrease the incidence of birth defects in Nevada and decrease the related morbidity, mortality, and disability that occurs when a child is born with a congenital or inherited disorder" (Nevada Department of Health and Human Services, 2006, para.1). The key activities of the Nevada Newborn Screening Program are to screen for metabolic, endocrine, hemoglobin, genetic or inherited conditions, provide short-term and long-term follow-up on these cases, provide case management, evaluation, treatment, and education to families and medical providers regarding the conditions identified at birth (Nevada Newborn Screening Practitioners' Manual, 2012).

The Nevada Newborn Screening Program identified a need for the review of sickle cell trait data in the State of Nevada because it has not been reviewed in the past. From this data, the Nevada Division of Public and Behavioral Health requested the development of some form of education to bring awareness to the potential health risks associated with SCT. Since this would be the first attempt at developing education for Nevada residents regarding SCT it was critical to perform a needs assessment in order to understand the existing knowledge related to SCT within the local communities, as well as to understand what the needs and preferences were for education. It was decided by the Newborn Screening program that a comprehensive educational campaign or program would be developed and would be based on this needs assessment (survey) administered to parents and medical providers throughout the State of Nevada. The present survey was the first documented assessment to evaluate existing statewide knowledge and needs regarding sickle cell trait in the State of Nevada.

The PRECEDE-PROCEED Model for Health Promotion Planning and Evaluation

The PRECEDE-PROCEED model is described as a “Community oriented participatory model for creating successful community health promotion interventions" (Berkowitz & Green, 2013, para.4). The PRECEDE-PROCEDE model is composed of eight phases or steps, and it begins with the final consequences and works backwards to the causes, and once the cause is known, an intervention can be designed (McKenzie, Neiger, & Thackeray, 2009). In the case of SCT in Nevada, the overall goal is to increase awareness of sickle cell trait and the potential health implications; however we wanted to first determine existing knowledge, and understand the best way to deliver education related to SCT to parents and medical providers. The PRECEDE phases (Phases 1-4) of the PRECEDE- PROCEED health planning model was used as a general context to determine existing knowledge and educational needs for sickle cell trait amongst parents and medical providers in the State of Nevada (Appendix E).

Purpose

This paper will detail the methods and results of a statewide needs assessment for the development of tailored sickle cell trait education. The purpose of this assessment is to determine existing knowledge and preferred educational approaches regarding sickle cell trait for parents and medical providers in the State of Nevada. The intended use of the results is to increase medical provider and parental knowledge and awareness of the potential health risks associated with sickle cell trait, and to give medical providers and parents of children born with sickle cell trait in the State of Nevada access to SCT educational resources.

The objectives of this research as conveyed in this professional paper are:

- To determine the existing knowledge and awareness of potential health risks associated with sickle cell trait among parents of children with sickle cell trait and medical providers in the State of Nevada.

- To determine the preferred educational approaches of parents and medical providers that can be used to develop tailored sickle cell trait education

Methods

Study Design

A cross-sectional survey design was used to obtain information from parents of children with sickle cell trait and medical providers on sickle cell trait education needs in the State of Nevada. Two key informant interviews were also conducted.

Survey Development

The surveys were developed between June 2013 and July 2013. Some of the survey questions were adapted from previous sickle cell disease awareness questionnaires (Boyd et al., 2005; Long, Thomas, Grubs, & Krishnamruti, 2011; Treadwell, McClough, & Vichinsky, 2006). Excluding demographic information, the surveys covered three main areas: general knowledge of sickle cell trait, newborn screening and sickle cell trait, and sickle cell trait education. Refer to appendix B for the parent and provider questionnaires.

Sampling and participants. The first survey was designed for parents whose children had been identified as carriers of sickle cell trait in the State of Nevada during the years of 2005 through 2012 in data provided by the newborn screening program. Participants were recruited from a database that was created through the Web Enabled Vital Records Registry System (WEVRRS), and matched with DMV records. The matched database was created through the Nevada Division of Public and Behavioral Health's (NDPBH) Office of Public Health Informatics and Epidemiology (OPHIE). From the matched DMV-WEVVRS database, 5,326 Nevada newborns were identified as variant trait carriers (including HbS, HbC, and Barts [alpha thalassemia]) during the years 2005 through 2012. Of this number 2,957 were sickle cell trait carriers (indicated as Hemoglobin FAS pattern in the database). From the 2,957 identified sickle cell traits, there were approximately 1,100 solid matches between the WEVRRS database and

DMV records that enabled the contact of parents of children born with sickle cell trait during the years 2005 to 2012. The eligibility of parent participants was based on the following criteria: parent of a child who has been identified as a carrier of sickle cell trait in the State of Nevada during the years 2005 through 2012, the parent is 18 years of age or older at time of the survey, and have a current home address within the State of Nevada at the time of the survey. The age appropriateness and readability of the parent surveys were evaluated prior to study implementation by external review.

The second survey was designed for medical providers who interpret newborn screening results in the State of Nevada. Participants were recruited through a contact list maintained at the NDPBH and the American Academy of Pediatrics (AAP) Nevada Chapter's contact list. Between the provider contact list from the NDPBH, and the contact list from the AAP Nevada Chapter there were approximately 900 physicians who interpret newborn screening results at the time of this assessment. The eligibility of medical providers was based on the following criteria: licensed medical practitioner in the State of Nevada (Physician), interprets newborn screening results either initial (hospital) or follow-up (primary care provider), and have a current physical address in the State of Nevada or e-mail address at the time of the survey.

A systematic random sample of 300 parents who have children born with sickle cell trait in the State of Nevada during the years 2005 through 2012 was taken from the matched parent contact database. The contact lists from the AAP and the provider lists at the NDPBH were combined. Then a systematic random sample of 300 providers was taken.

Survey Implementation

The Institutional Review Board (IRB) at the University of Nevada, Reno approved all components of this study prior to implementation. IRB number 2014S002 was assigned to this project (Appendix A). The survey period lasted from July 21, 2013 to November 21, 2013. The two surveys were distributed over three consecutive periods (Figure 2). The first survey period lasted from July 21, 2013 to August 21,

2013. The second survey period lasted from September 21, 2013 to October 21, 2013, and the last survey period occurred during October 21, 2013 to November 21, 2013. The reason why there was a month delay in between the first and second wave of surveys was due to approvals needed to send amended survey cover letters for parents with the survey link online through the NDPBH; these approvals were in process during this time frame.

The survey was mailed to the parent's home address with a pre-paid postage return envelope included. During the first survey wave, parents were sent a paper and pencil version of the survey. During the second and the third survey waves, parents were sent a letter along with a link to an online survey option through the online survey manager Survey Monkey; however they still had the option to submit the survey via standard mail. The survey was sent to providers using standard mail and e-mail. The e-mailed surveys were sent through Survey Monkey. Surveys were sent through e-mail to the AAP providers because there was a reliable e-mail contact list for the chapter.

A cover letter was sent to survey participants along with the survey. The content of the cover letter included: an overview of the purpose of the research study, a description of how the respondent's names were obtained, emphasis on the confidentiality of results, and contact information for the research team. By submitting the survey back to investigators, informed consent was implied. If participants had not replied to the survey by the stated deadline, a second questionnaire was sent during the second survey wave, if no response was received during this timeframe, a postcard along with a third questionnaire was sent to non-respondents during the third-survey wave indicating the last chance to participate.

Key Informant Interviews

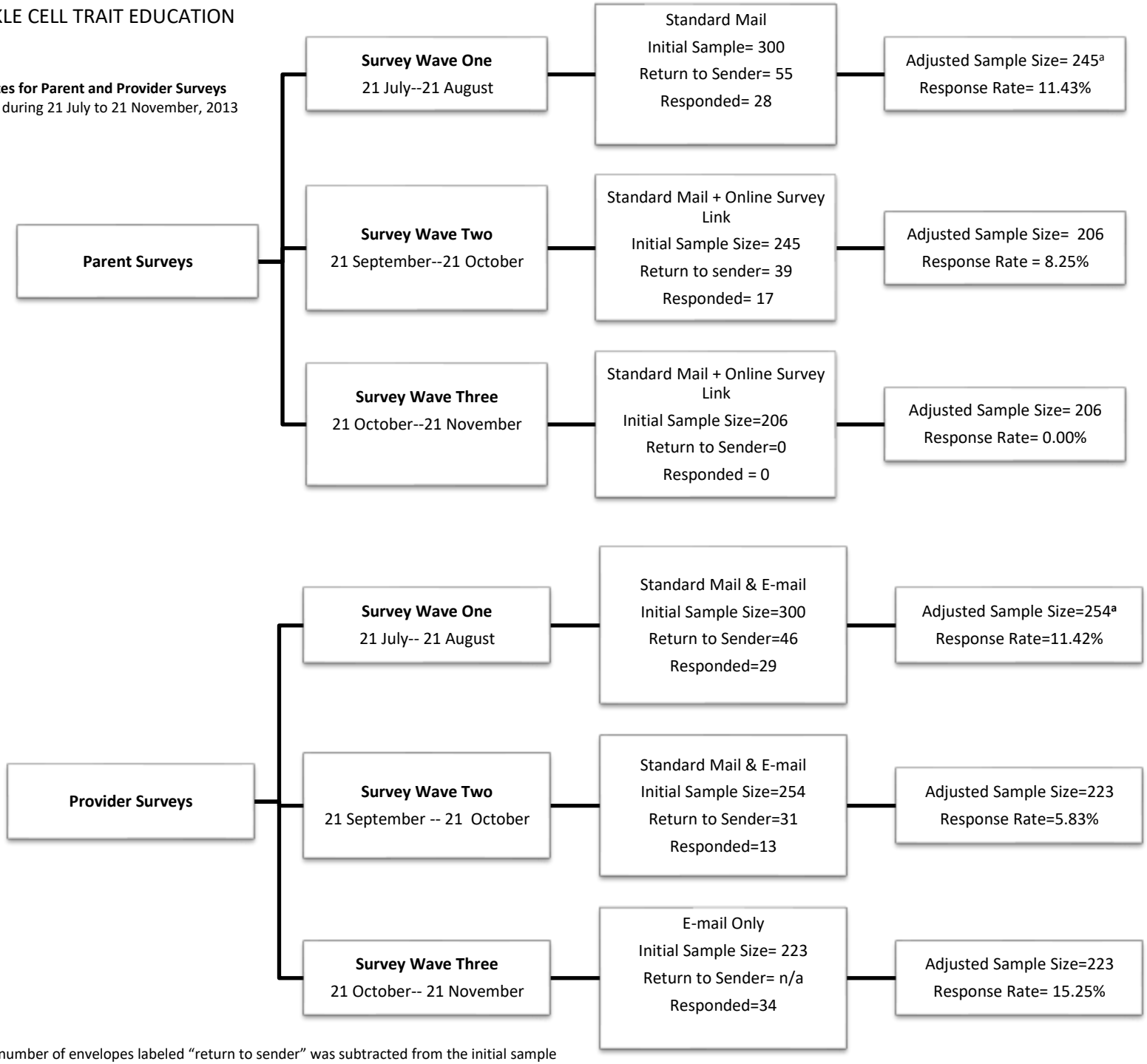
The key informant interviews occurred in August of 2013. The eligibility criteria for the key informant candidates were as follows: Licensed Practitioner (Physician or Registered Nurse); specializes in hematology, medical genetics, or newborn screening; and have at least five years of clinical

experience with hemoglobinopathies (specifically sickle cell disease or sickle cell trait) by the interview date. Two practicing physicians in the State of Nevada fit the eligibility criteria, and were interviewed to gain a well-rounded perspective on issues regarding sickle cell trait in the State of Nevada. These physicians were selected for the key informant interviews based on their research and work in sickle cell anemia and sickle cell trait as well as their noted reputation as expert clinicians within the community.

Analyses

Quantitative survey data were analyzed using Statistical Package Software (SPSS) version 20 for Windows. Standard descriptive statistics such as frequency distributions (numbers and percentages) were performed on all close-ended survey questions. All of the percentages displayed are the valid percentages, and excludes people who did not answer the specific question. Chi-square tests were performed utilizing Microsoft Excel to compare key demographic variables among parent responders and parent non-responders, as well as early provider responders and late provider responders. This was done to address the potential for unit non-response bias. There was an open-ended comments section in the survey for parents and providers. Qualitative survey data were analyzed for major themes. Each open-ended response was coded into specific categories. The detailed results of the open-ended questions can be found in appendix D. Key informant interviews were also summarized for major themes in this paper.

Figure 2: Response Rates for Parent and Provider Surveys
Survey Waves 1, 2, & 3 during 21 July to 21 November, 2013



a --The number of envelopes labeled "return to sender" was subtracted from the initial sample size to obtain the adjusted sample size during each survey wave for both parent and provider

Results

Participants Demographics and Response Rates

Of the six hundred surveys mailed (parents and providers combined), one hundred and twenty-one surveys (n=121) were returned and eligible for analyses. The response rate for parent surveys was 22%, and the provider survey response rate was 34%. There were issues with a high non-response rate for parents (78%) and providers (66%). This was partially related to a high volume of envelopes that were received back stating “return to sender” from both parent and provider surveys. The number of surveys received back as “return to sender” was deducted from the initial sample size. The response rates were also calculated for each survey wave (1, 2, and 3). It was important to calculate the response rates separately for comparison purposes (Figure 2). The first wave of surveys for the parents was strictly done by standard mail for both sending and returning the surveys. However, the second wave of surveys for the parents included a modified cover letter with a link to the survey online. It was essential to make this distinction in order specify what method of communication was most effective in terms of reaching our specific sample. The highest response rates were for parents via standard mail without the online survey link during the first survey wave (11%); and the highest response rate for providers were by e-mail (15%) during the third survey wave (Figure 2). It is also worth mentioning that outside assistance was received in distributing provider surveys during the third survey wave.

Due to the high non-response rates it was essential to address the potential of unit non-response bias in the sample. For the parents, this was accomplished by comparing selected demographic variables among those participants who responded and those who did not respond. For the parent surveys, demographic data of the non-respondents was available through the DMV-WEVRRS matched database. On average the parent participants who responded and those who did not respond were of similar age. Parents who responded were a mean age of 31.84 years old, standard deviation (SD) = 6.33; while non-responder parents were a mean age of 32.02 years old, SD=6.89. Likewise, the

racial distribution of the parent respondents and parent non-respondents was similar. Demographic differences among parent responders and parent non-responders by race was examined using a chi-square test with a level of $p<0.05$ considered as significant. American Indian/Alaskan Natives was excluded from this analysis due to the small number of responses. There was no significance found between race and response for parents ($p > 0$ .05).

In terms of addressing non-response bias for providers, late responders were compared to early responders. In this case late responders were used as a proxy for non-responders because responders were defined as those who sent their surveys back within the first survey wave (n=29). Late responders were defined as those who sent their surveys back in the second and third survey waves (n=47). The demographic variables that were compared included: race and gender, and the number of years the physician had practiced medicine in the State of Nevada up to the survey date. On all three variables, differences between the early and late responders was examined using a chi-square test with a level of $p <0.05$ considered significant. For race, the American Indian/Alaskan Natives sub-category was excluded from this analysis due to a small number of responses. There was no significance found between race and response between physician early responders and late responders ($p>0.05$). Similarly, there were no significant differences for the variable of gender between early responding physicians and late responding physicians, ($p>0.05$). Statistically significant differences were found for the number of years a physician had practiced medicine in the State of Nevada and survey response ($p<0.05$). The percentages demonstrated that late respondents were more likely to have practiced medicine in Nevada for less than one year (16 % for late responders, and 4% of early responders). While early respondents were more likely to have practiced medicine in the State of Nevada for a period of either one to five years or five years and greater (96% of early responders , and 84 % of late responders).

Table 1 outlines the demographic characteristics of both the parent and provider survey respondents on the selected variables of age, gender, ethnicity, race, and the number of years they have

resided in the State of Nevada. Only parents were asked the total number of children they have, and the number of children they have with sickle cell trait; this information is also displayed in table 1.

Table 1: Selected Demographics of Parents and Providers

	Parents		Providers	
Age	**N**	**(%)[a]**	**N**	**(%)**
18-24	2	(4.40)	2	(2.70)
25-34	22	(47.80)	32	(43.24)
35-44	18	(39.10)	15	(20.27)
45-54	2	(4.30)	13	(17.57)
55 or older	0	(4.40)	12	(16.22)
Total	45	100.00	74†	100.00
Gender	**N**	**(%)**	**N**	**(%)**
Male	0	(0.00)	32	(43.24)
Female[b]	44	(100.00)	42	(56.76)
Total	44*	100.00	74†	100.00
Ethnicity	**N**	**(%)**	**N**	**(%)**
Hispanic	11	(25.00)	3	(4.05)
Not Hispanic	33	(75.00)	71	(95.95)
Total	44*	100.00	74†	100.00
Race	**N**	**(%)**	**N**	**(%)**
White	13	(30.23)	41	(58.57)
Black/African American	19	(44.19)	6	(8.57)
Asian	3	(6.98)	17	(24.29)
Native Hawaiian or Other Pacific Islander	0	(0.00)	0	(0.00)
American Indian or Alaskan Native	0	(0.00)	1	(1.43)
Other	8	(18.60)	5	(7.14)
Total	43†	100.0	70‡	100.0

a-- The valid percent for each variable is reported in this table.
b-- Parent participants had 100% female respondents, due to the individuals name on record (usually the mother's name) at the NBS program.
*-- Missing information for 1 respondent
† -- Missing information for 2 respondents
‡ --Missing information for 6 respondents

Table 1 Continued : Selected Demographics of Parents and Providers				
	Parents		Providers	
Number of Years Resided in Nevada	**N**	**(%) [a]**	**N**	**(%)**
< 1 year	1	(2.27)	9	(12.17)
1-5 years	6	(13.64)	25	(33.78)
>5 years	37	(84.09)	40	(54.05)
Total	44*	100.00	74[†]	100.0
Children [‡]	**Average**	**Standard Deviation**	-	-
Number of Children	2.12[c]	1.21	-	-
Number of Children with SCT	1.22	0.67	-	-
Total	43[†]			

a-- The valid percent for each variable is reported in this table.
c --These numbers are averages
†-- Missing information for 2 respondents
‡--Only parents were asked this survey question

General Knowledge of Sickle Cell Trait

The parent survey and the provider surveys covered the same overall areas, but had different questions in the section regarding general knowledge of sickle cell trait. General knowledge questions intended to assess the participant's baseline understanding of sickle cell trait, and also to determine what information may be useful to include in the development of sickle cell trait educational materials.

General knowledge: parent survey. On a five-point Likert scale, participants were asked to rate their knowledge of genetics, with one being poor and five being excellent. The majority of the participants (40%) rated their knowledge of genetics as good; only 6.7% rated their knowledge of genetics as poor. Participants were also asked if they had ever heard of the term sickle cell trait, and 93.3% answered that they had heard of the term sickle cell trait, 4.4% had not, and 2.2% were not sure. Participants were also assessed on their knowledge of the difference between sickle cell trait and sickle cell disease. Six-percent of participants believed that there is no difference between sickle cell trait and sickle cell disease, while 93% of participants believed there was a difference. Thirty-one percent of

participants believed that sickle cell trait can turn into sickle cell anemia, while 68% of participants did not believe this statement. To gauge knowledge on potential health implications as well as SCT misconceptions, participants were asked to select true statements about sickle cell trait (Table 2). Eighteen-percent of parent participants believed that sickle cell trait would never lead to health problems, 23% of parent participants believed that is was strictly a condition that affects African-Americans, 4% believed that all of the statements listed in the survey were correct, and 53% of respondents selected that none of the statements listed were true (all of the statements listed were incorrect).

Table 2: Survey Question Assessing General Knowledge of Sickle Cell Trait

Question: **Which of the following statements is true regarding sickle cell trait?**	**Parents**		**Providers**	
Answer Options:	**N**	**(%)**	**N**	**(%)**
Sickle cell trait will never lead to any health problems.	8	(18.60)	12	(15.79)
Sickle cell trait is a more severe form of sickle cell anemia.	0	(0.00)	0	(0.00)
Only African-Americans can inherit sickle cell trait.	10	(23.26)	1	(1.32)
All of the above statements are true.	2	(4.65)	0	(0.00)
None of the above statements are true.	24	(53.49)	64	(82.29)
Total (N)	43†		76	

†-- Missing information for 2 respondents

General knowledge: provider survey. Physician respondents were also asked to provide a rating of their general knowledge of genetics, 52.6% rated their knowledge as good, 2.6% as excellent, and 29% as fair. Providers were also asked similar questions to the parents that gauged their knowledge on the potential health implications of SCT, as well as SCT misconceptions. Fifteen-percent of the providers surveyed believed that sickle cell trait is a benign condition that will never lead to health

problems, one respondent (1.3%) believed that African-Americans were solely affected by sickle cell trait, and the majority of participants (82.9%) selected that none of the statements regarding SCT were correct (Table 2).

Newborn Screening and Sickle Cell Trait

The questions in the second part of the survey had a different focus for parents and providers. For parents the intention was to assess familiarity with the newborn screening process, attitudes and beliefs regarding genetic screening, their main sources of obtaining medical information, and their perceived resources for sickle cell trait in their local community. For medical providers the questions focused on their perceived role of the physician in the screening and follow-up processes, general attitudes towards genetic screening, clinical familiarity with sickle cell trait, and their perceptions on why poor medical follow-up occurs for individuals with sickle cell trait.

Newborn screening and sickle cell trait: parent survey. The majority of parent respondents were aware of the newborn screening process for sickle cell trait (93.3%). Some respondents were also aware that adults can receive a lab test to find out their SCT status (57.8%). Parents were also asked to rate their attitudes towards newborn screening in general (positive or negative), and the majority of respondents (64.5 %) had a positive attitude towards genetic screening. Participants were asked if they had at least one doctor who was considered to be their child's primary doctor or pediatrician, and the majority (93.3%) did, only 6.7% of respondents did not. Seventy-one percent of respondents received most of their health related information from a healthcare professional, and 26.7% received their information from the internet; parents were also afforded the option to provide an open response to this question (appendix D). Parents were also asked if they had received any educational information related to sickle cell trait within the past five years, and were also asked on a five-point Likert scale to rate their level of satisfaction with the information they had received with one being very dissatisfied and five being very satisfied. Thirty-seven-percent of participants reported they had received

educational information, 53.3% of participants had not received any information, and 8.9% were not sure if they had received any information in the past five years. Participants who had received information rated their satisfaction as satisfied, 37.8% (very satisfied/satisfied), neither satisfied or dissatisfied was 31.1%, and dissatisfied 31.1% (very dissatisfied/dissatisfied). The majority of respondents (68.9%) did not feel as though they had adequate resources in their local community for their child's sickle cell trait such as genetic counseling, education, and support.

Newborn screening and sickle cell trait: provider survey. Providers were also asked to rate their general attitude towards the newborn screening process (positive or negative), and 94.7% rated a positive attitude towards genetic screening. To gauge the familiarity of providers in the State of Nevada with sickle cell trait health issues, providers were asked to rate on a five-point Likert scale how often they saw patients with sickle cell trait, with one being not all and five being very often. A majority of physician respondents did not see patients with sickle cell trait, (not often = 63.5%; often=36.5%). Physicians were also asked to rate on a five-point Likert scale their perceived importance of the physician's role in referring patients with sickle cell trait and their families to follow-up services (such as trait counseling), with one being not at all important and five being very important. The majority of respondents believed that the physician's role is important, (96 %), while 4% believed that the physician's role is not important in this process. Lastly, physicians were provided with a list of statements and they were asked to select which statements best described the reasons for poor medical follow-up for individuals with sickle cell trait; physicians were also given the option to provide an open response to this question (appendix D). The majority of responders (68%) selected the statement, "Primary healthcare providers focus on benign nature of trait, not on future implications" as a main reason for the poor follow-up of individuals with sickle cell trait. There was also a high percentage for the statements, "Parents don't understand the trait, and do not think it affects them" (58.3%), and

"There is a lack of education in the community, and most people have no idea about sickle cell trait" (52.8%).

Sickle Cell Trait Education

The final section of the survey was the same for both parents and medical providers; therefore the results are displayed here simultaneously. The questions assessed the participant's perceived need of sickle cell trait education and resources as well as the preferred methods of SCT education delivery. The majority of parents (93.3%) agreed that they would like more opportunities to learn about sickle cell trait; and 86.7% wanted access to general resources and support services regarding sickle cell trait. Likewise, 76.3% of providers agreed that they would like more opportunities to learn about sickle cell trait, and 80.3% wanted access to more resources regarding sickle cell trait. Additionally, participants were asked what topics would interest them in an educational program related to sickle cell trait. Participants were provided with a list of topics and among parents, the two most selected topics were: potential health problems associated with sickle cell trait (75.6%), and preventive measures for potential health problems associated with sickle cell trait (80%). Among providers the two most selected topics were the same as the parent respondents at (76.3%), and (76.3%) respectively (Table 3).

Table 3: Survey Question Assessing Topic Preferences for Sickle Cell Trait Education

Question: If an educational program were to be developed addressing sickle cell trait, what topics related to sickle cell trait would you be interested in?	Parents				Providers			
Answer Options:	**Little or No Interest**	**Some Interest**	**Strong Interest**	**Total**	**Little or No Interest**	**Some Interest**	**Strong Interest**	**Total**
	N (%)	**N (%)**	**N (%)**	**N (%)**	**N (%)**	**N (%)**	**N (%)**	**N (%)**
Genetics of sickle cell trait.	2(4.44)	15(33.33)	28(62.22)	45(100.00)	8(10.67)	40(53.33)	27(36.00)	76(100.00)
Follow-up services available for sickle cell trait.	2 (4.44)	17(37.78)	26(57.78)	45(100.00)	0(0.00)	20(26.32)	56(73.68)	76 (100.00)
Trait counseling.	7(15.56)	15(33.33)	23(51.11)	45(100.00)	2(2.63)	23(30.26)	51 (67.11)	76 (100.00)
Potential health problems associated with sickle cell trait.	2(4.44)	9(20.00)	34(75.56)	45(100.00)	2 (2.63)	16(21.05)	58(76.32)	76 (100.00)
Measures to prevent for potential health problems associated with sickle cell trait.	1(2.22)	8(17.78)	36(80.00)	45(100.00)	2 (2.63)	16(21.05)	58(76.32)	76 (100.00)

Participants were also asked about their preferred methods of delivery for SCT education. Participants were provided a list describing a variety of delivery methods and asked to select their level of interest in each method. Amongst parents, the most selected delivery method was a website dedicated to sickle cell trait education and resources (77.8%); this was also the most selected educational delivery method amongst medical providers at 73.3% (Table 4).

Table 4: Survey Question Assessing Format/Delivery Preferences for Sickle Cell Trait Education

Question: Please indicate the format/delivery that you have interest in for an educational program about sickle cell trait.	Parents				Providers			
Answer Options	**Little or No Interest**	**Some Interest**	**Strong Interest**	**Total**	**Little or No Interest**	**Some Interest**	**Strong Interest**	**Total**
	N (%)	**N (%)**	**N (%)**	**N (%)**	**N (%)**	**N (%)**	**N (%)**	**N (%)**
Mail out written information or pamphlet.	3(6.67)	14(31.11)	28(62.22)	45(100.00)	21(28.38)	30(40.54)	23(31.08)	76(100.00)
Informational Community Meetings.	18(40.00)	17(37.78)	10 (22.22)	45(100.00)	35(46.67)	30(40.00)	10(13.33)	76(100.00)
Publicize on Television.	12(26.67)	22(48.89)	11 (24.44)	45(100.00)	29(39.19)	31(41.89)	14(18.92)	76(100.00)
Publicize on radio.	21 (46.67)	18(40.00)	6 (13.33)	45(100.00)	33(44.59)	29(39.19)	12(16.22)	76(100.00)
Distribute a video or CD about Sickle Cell Trait.	8(17.78)	17(37.78)	20 (44.44)	45(100.00)	23(31.08)	37(50.00)	14(18.92)	76(100.00)
Website dedicated to sickle cell trait education and resources.	2(4.44)	8 (17.78)	35(77.78)	45(100.00)	2(2.67)	18(24.00)	55(73.33)	76(100.00)
Community-based events.	13(28.89)	15 (33.33)	17 (37.78)	45 (100.00)	20(27.40)	30(41.10)	23(31.51)	76(100.00)

Comments and Concerns Regarding a Sickle Cell Trait Education Program

Both parent and provider participants were asked if they had any comments or concerns regarding a sickle cell trait education program in an open-ended comment box. A total of fourteen participants provided a comment (eight parents and six providers). The qualitative analysis was done separately for parents and providers (appendix D).

Parent survey comments. A common concern among parents who provided comments was the lack of educational and medical resources dedicated to sickle cell trait and sickle cell disease in the State of Nevada. Some parents believed that sickle cell trait education was critical at the time of birth, while some commented that follow-up education throughout a child's early years was essential. For example, one parent stated, "So far our son has not had any problems, but I am interested in more information to help as he grows to understand the trait that he carries."

Another issue that was found in the qualitative analysis of the parent responses was that parents are themselves unclear or unsure of their own trait status, as well as the differences between sickle cell trait and sickle cell disease. An example statement provided by one parent was, "I would like to know what the cut-off number is since I was told I was on the borderline, and I need to know if I have the trait or the disease."

Provider survey comments. A common theme found among provider comments were specific suggestions with regard to the implementation of a sickle cell trait education program. Some providers suggested that online materials would be the most effective means to implement education, there were also suggestions to put handouts, resources, and other related information on a website that could be easily accessed and printed. Some providers believed a more collaborative effort was needed within the community in terms of sickle cell trait education. For example, one provider stated, "There has to be a concerted effort to [sickle cell trait] education from the community, physicians, and families; and collaborations need to be formed with concerned groups."

Another concern that came from the qualitative analysis of the provider surveys was that of trait status awareness and screening. One provider stated, "Clinicians need to know that every individual should be aware of their status as everyone is screened at birth." The final theme that emerged from the qualitative analysis was that of program costs. Some physicians were concerned whether an

educational program dedicated to sickle cell trait would be expensive or if an educational program would even be covered by insurance.

Key Informant Interviews

Two practicing physicians licensed in the State of Nevada were interviewed regarding community and state needs for sickle cell trait education. The key informants were asked the same interview questions (appendix C). Qualitative analyses were performed on the interview responses and similar responses were grouped into categories. The themes that emerged from the interview responses are described in this narrative. To protect the identity of key informants, they will be referred to as KI one and KI two in this narrative.

One common theme that emerged from the interviews was the need for increased awareness related to sickle cell trait and the potential health implications associated with it. KI two is a pediatric hematologist, and they mentioned that some of their patients with sickle cell trait have renal related issues, and their parents are never sure what to do because they were not aware that there was any health implications associated with sickle cell trait. KI one was interested in the research aspects related to sickle cell trait, and truly believes that any interventions related to sickle cell trait can be considered preventive measures.

Another key issue that emerged from the interviews was a critical lack of resources in the State of Nevada in relation to sickle cell trait and sickle cell disease. KI one noted that there are insufficient resources for families who have children with sickle cell trait; a key resource being genetic counseling, and genetic specialists in the state.

Both key informants recommended that an educational program for sickle cell trait should not only target parents at the time of newborn screening, but there needs to be a continuous educational process that follows a child throughout their childhood and teenage years, with a dialogue that involves medical providers. KI one recommended implementing educational curriculum in the schools as well as

implementing a sickle cell trait awareness campaign in local churches. KI two also recommended implementing a sickle cell trait awareness campaign that was centered in local faith-based organizations.

Both key informants believed that the internet was an important medium for reaching out to the community for sickle cell trait education. KI two mentioned that there are many sickle cell disease and sickle cell trait support groups on social media sites, and these may be important resources. KI two also mentioned that most of their patients prefer to access educational materials easily, and suggested that a website with resources for sickle cell trait may be an effective method to reach Nevada's population.

Discussion

This was the first documented assessment of existing knowledge and needs regarding sickle cell trait in the State of Nevada. Overall, the results of the surveys and key informant interviews indicate a significant need for dedicated resources related to SCT in the state. These resources include genetic counseling and educational materials that can be accessed by parents, medical providers, and the general public. As the study results demonstrate, the majority of parents surveyed did not believe that they had adequate support services or educational information for their child's SCT. Moreover, the majority of parents surveyed indicated that they received most of their health information from a medical provider, and that they a have a primary pediatrician for their child. This physician-patient relationship suggests key opportunities where education and awareness regarding SCT can occur, and as our study results convey, physicians in our state see their role in the educational process as fundamental.

In terms of existing knowledge of SCT for parents, our data is comparable to other studies assessing community level knowledge of SCT and sickle cell disease. The majority of the parents surveyed had adequate knowledge of SCT, but this could also be because they were a very specific sample: they are parents of children with SCT. Other surveys have examined a more generalized

population, such as people with or without SCT and SCD (Boyd et al.; Long, Thomas, Grubs, Gettig, & Krishnamruti, 2011; Treadwell, McClough, & Vichinsky). Additionally, we assessed parent's knowledge on the difference between SCT and sickle cell disease, and the majority of parents who responded knew the difference. Other sickle cell disease and sickle cell awareness surveys specifically noted that they did not assess if participants understood this difference (Long, Thomas, Grubs, Gettig, & Krishnamruti, 2011). Generally, medical providers had an adequate understanding of SCT, but a majority of responders indicated that they never or rarely saw patients with SCT. This could potentially be an indication of the demographics of the population areas that the physician respondents serve, or it can also be an indication of a lack of resource knowledge or utilization amongst individuals with SCT.

The study findings also highlight a need to increase awareness regarding common misconceptions about SCT. A considerable percentage of parent respondents believed that SCT solely affects African-Americans. Furthermore, a great percentage of medical provider and parent respondents believed that SCT is completely benign, and has no health implications. Despite these responses, the majority of participants, both parents and providers, were interested in gaining more information about the potential health problems associated with SCT, and understanding preventive measures for these potential health problems. As with previous study findings, our study results found that there are many misconceptions surrounding SCT and that health care providers may be the critical link that can help to dispel these misconceptions. As our study results revealed, the majority of parents received most of their SCT information from their medical provider. A 2009 study by Acharya, Walsh, & Friedman-Ross, examined knowledge, attitudes, and beliefs about SCT sources of information from parents with SCT and parents of a child with either SCD or SCT. The study findings revealed that the majority of parents with an affected child cited pediatricians or an SCD clinic as the common source of information. Additionally, participants in this 2009 study had significant misconceptions regarding their own carrier status,

reproductive implications, and noted that professional counseling for SCT or SCD was rare (Acharya, Walsh, & Friedman-Ross, 2009).

In terms of educational delivery needs, both parent and provider respondents preferred an educational delivery method that was online, such as a website with dedicated resources to SCT. The key informant interviews reinforced the internet as a consideration in the development of SCT educational materials because both key informants believed that the internet was an important medium for reaching out to the general community for SCT education.

In addition to being the first survey to assess knowledge and needs regarding SCT in the State of Nevada, this study adds to the very limited information available regarding state or community level SCT knowledge in the current literature. Furthermore, this study assessed medical providers and parents statewide which are a broad sample compared to other sickle cell awareness surveys. Previous studies have usually been limited to men or women living in specific neighborhoods or of specific racial and ethnic backgrounds. The inclusion of medical providers is a key strength of this study because of the dialogue that has been ongoing in the medical community regarding SCT as a medical condition or a benign carrier state. Despite extensive evidence that supports the morbidity and rare, but documented, mortality associated with SCT, there is yet to be consensus on this issue. Therefore, it is essential that there is at least some awareness brought to SCT, especially within the medical community.

Another key strength of this study is the fact that the surveys utilized were built upon previous sickle cell awareness questionnaires. As Boyd et al. mentioned in their 2005 study that specifically assessed community knowledge of sickle cell disease in African-American women, “To our knowledge, no standard set of questions have been used to assess knowledge about SCD. Future studies would benefit from the inclusion of similar questions to determine the reliability of such questions in assessing knowledge of SCD” (p. 66). While our study pertains to SCT, not SCD, it does add to the general foundation of increasing evidence in the area of sickle cell community knowledge.

Study Limitations and Suggestions for Improvement

A significant limitation in this study was the low response rate amongst parents and providers. It is generally acknowledged that it is difficult to gain a high response rate from physicians as a professional group (Thorpe et al., 2008). The provider response rate was 34%, which may be considered an adequate response rate for physicians based on the current literature in survey research. A 2011 research study surveyed pediatricians in three specialty sections of the American Academy of Pediatrics regarding SCT screening of college athletes per NCAA recommendations, this study yielded a response rate of 44%, (254 of 574 eligible respondents) (Koopmans et al.). The response rate of 22% for parents was quite low, but was also anticipated considering the use of DMV addresses for the parent surveys, and the highly transient nature of Nevada's population. However, the main issue with a high non-response rate is the increased risk of introducing non-response bias. While analyses were performed to compare demographic characteristics among responders and non-responders, these are limited because we only compared within the sample and only compared select demographic characteristics. Furthermore, a small sample size as a result of the high non-response (n=121), and a very specific sample limits the generalizability of the results to other populations.

Our survey results highlight that e-mail was the best method to reach medical providers when compared to standard mail. For parents, we did not have the option to send the survey via e-mail; however during the second and third survey waves we sent a modified cover letter with a link to the survey online. Nonetheless, we do not have enough data to compare the best methods to reach parents from our sample. Recommendations for improvement for the low response rate when opting to use the mailed survey may be to send surveys through a tailored design method (TDM), also known as the Dillman Approach. The Dillman Approach involves five key elements to achieving high response rates and is often considered the gold standard approach to mailed surveys (Thorpe et al., 2008). The five steps of the Dillman Approach include: (1) a respondent friendly questionnaire, (2) use of four contacts

by first class mail with an additional special contact (i.e. certified mail or telephone call), (3) use of real first class mail stamps, (4) personalized correspondence, (5) a token or financial incentive (Thorpe et al. 2008). While most of the survey methodology in our study closely mirrored the Dillman Approach of survey implementation, there were some steps that were simply not feasible due to the nature of the project and budgetary constraints. For example, we were unable to initiate a fourth contact or a special contact due to time issues and manpower. Furthermore, we were unable to offer a financial incentive due to budget constraints. Nevertheless, these may be important considerations in terms of reaching out to the population addressed in our study in order to obtain a higher response rate in future studies.

Another limitation in terms of the parent surveys was related to gender. In the DMV-WEVVRS matched database, the mother's name is registered as the primary contact; therefore the surveys were addressed to the mother of the household. As a result, our sample of parents was 100 % female, and we were unable to generalize the "parent" results to fathers of children with SCT. A recommendation for improvement on this limitation, when reaching out to the families who have the mother's contact information on the Newborn Screening file, may be to address the letter to the family. For example, address the envelope "to the Jones Family" or "to the parents of" the child who is being inquired about, in order not to limit who answers the questionnaire. Another recommendation could be to ensure that the cover letter clearly outlines that the either the father or mother of the child can answer the survey.

Another limitation related to the parent surveys was the language appropriateness of the surveys and cover letters for individuals who do not read or speak English as their primary language. Nevada has a growing Hispanic/Latino population, and according to the 2012 US Census Bureau the Hispanic/Latino population makes up 27.3% of Nevada's population (US Census Bureau, 2012). Furthermore, SCT has been documented in individuals of Latino heritage (Vichinsky, 2012). A 2010 statewide needs assessment by the University of Nevada, Reno Cooperative Extension that focused on Latino population needs for Nevada indicated that the majority of their survey respondents (49.3%)

spoke Spanish at home, only 7.8 % of the respondents from that survey spoke English at home (Skelly, Singletary, Angle, & Sepúlveda-Pulvirenti., 2010). Our cover letters and surveys were written in English only. We were unable to provide a cover letter and survey in Spanish due to time and budget constraints, however this is an essential consideration in future studies that will address sickle cell trait in Nevada. The inability to provide letters in both English and Spanish may have also contributed to the lower response rate.

A final limitation of this study was the depth of some the survey items in terms of measuring precise knowledge of a particular subject. One example was the question pertaining to the individual's knowledge of genetics. This specific question is based more on self-perception of genetics knowledge rather than a concrete measure. It would be important to review other survey items in existing literature regarding knowledge of genetics so that future questions assessing genetic knowledge can provide a more concrete measure. Some examples may include: having participants defining what a gene is, or describing how SCT is inherited.

Study Implications

Our study results indicate a significant need for increased education and awareness for SCT, and the health issues associated with SCT in the State of Nevada. Many parents surveyed believed that they have received either no information regarding SCT or inadequate information. While the Internet can be a key method to reach out to individuals with SCT and their families, utilizing community organizations for outreach may be another practical option. As the key informant interviews suggested, reaching out through schools or faith-based organizations may be an effective method for individuals with SCT and their families. In fact, one of the Healthy People 2020 objectives that pertain to blood disorders is to "Increase the proportion of community based organizations (CBOs) that provide outreach and awareness campaigns for hemoglobinopathies" (US Department of Health and Human Services, 2013, BDBS-9). Implementing a health education and awareness program regarding SCT through a

combination of internet-based and local community organizations could put Nevada on the path to addressing the specific needs of people with SCT.

An important theme that was highlighted in the results of our research study, that has also been a recurrent issue in the literature regarding SCT awareness, is the need for a continuing dialogue regarding SCT throughout a person's lifespan. At this point, most of the education regarding SCT is inconsistent and is usually done on a need to know basis, or only when it is considered necessary. For example, education may be done at birth during the newborn screening process or when an individual is screened for college athletics. The lack of consistency regarding sickle cell education and follow-up is a key controversy regarding counseling and SCT; because over time people will forget or become unaware of their SCT status. According to Grant et al., (2011), "Information about SCT is not a one-time message from care providers; it must be repeated at various stages of life so that people have the information at times when it is most useful to them" (p. S438). This is an important practical implication because it may be a key focus of how medical providers need to approach patients with SCT. This may also imply that a broader range of physicians will need to have knowledge regarding SCT, not only hematological specialists and pediatricians.

Other suggestions for education have been to implement SCT awareness programs in schools. This suggestion may help to reach a more generalized population as well as keep individuals informed of SCT in a more longitudinal timeframe (e.g. from early school years throughout high school). Some have suggested implementing SCT awareness gradually throughout a child's years in school so that they will have the continual knowledge as they get older. Aggarwal (2011) outlined an SCT educational curriculum that is based in a variety of textbooks and lectures, and ranges from topics such as biology and genetics, geography, mathematics, computer sciences, history, sociology, health sciences, and physical education. According to Aggarwal, "When the students relate the study material being applicable to themselves or

their fellow students, they are more apt to generate interest in understanding the issue, and thus understanding the study material" (2011, p. 3).

In addition to increasing awareness to SCT in schools among students, it is equally important to address SCT awareness amongst educators, school administrators, and supportive staff such as coaches and school nurses. Staff should be aware of the potential health implications associated with SCT and the proper precautions to take to avoid adverse events. The Centers for Disease Control and Prevention have created a booklet for educators with guidelines to support children with sickle cell disease; this booklet does touch on some related SCT information (Centers for Disease Control and Prevention, n.d.). There are currently limited guidelines for educators, and supportive school staff dedicated exclusively to sickle cell trait, however recommendations are in place. In fact, the National Athletic Trainer's Association (NATA) recommends that all staff and students in educational establishments are educated about sickle cell trait. In their consensus statement titled, *Sickle Cell Trait and The Athlete*, the organization states, "Irrespective of screening, [educational] institution [s] should educate staff, coaches, and athletes on the potentially lethal nature of this condition [SCT]" (n.d., para. 5). Other organizations such as the NCAA provides educational information and resources specific for athletes and coaches about sickle cell trait and the information provides coaches general strategies for working with sickle cell trait athletes. Furthermore, staff should have emergency plans outlined in the event of an adverse situation associated with a student who has sickle cell trait; there are already standard guidelines in place by NATA and the NCAA that educational institutions can adopt (NATA, n.d. & NCAA, n.d.).

While increasing education and awareness for SCT are essential tasks, there are many key points to keep in mind while developing an educational program that could be tailored for SCT. In 2009, the Centers for Disease Control and Prevention (CDC) hosted a meeting on SCT which gathered representatives from the National Institutes of Health (NIH), the Sickle Cell Disease Association of

America (SCDAA), the Health Resources Services Administration (HRSA), and other key stakeholders. The main objective of this meeting was to discuss the public health implications of SCT, and one of the main topics addressed was communication, education, and awareness of SCT. Many suggestions were provided during the meeting, particularly suggestions of ways to implement community awareness and health education initiatives (Grant et al., 2011), and some of these suggestions are outlined in table 5.

Table 5: Suggested activities (selected) to move community awareness and health education regarding sickle cell trait forward. From the 2009 CDC Meeting on Sickle Cell Trait (Grant et al., 2011, p. S348).
Health education efforts should capitalize on existing resources that have been created by existing organizations, and/or funded by federal agencies such as HRSA.
Education and awareness about risks associated with SCT targeted towards policymakers, general public, media, and health providers, especially primary care providers, coaches, and employers and addressing needs of various populations should be improved.
Incorporating broad media sources and community institutions including faith-based organizations, schools, day care, and other community institutions should be developed.
Health education efforts should address variability in health literacy, cultural and linguistic diversity, reproductive decision-making, as well as adverse health outcomes for individuals.
Health education efforts should avoid increasing stigma and unduly worrying parents, athletes, and prospective parents.
Uniform consistent SCT messages should originate from authoritative public health sources such as the CDC, HRSA, NIH, and others
Programs are needed to support public understanding of genetic information. Participants suggested that health education messages may need to be tailored for SCT.

The suggestions provided at the 2009 CDC meeting can be a useful guide in developing SCT education and awareness programs. The recommendations also provide insight to the considerable scope of work that is needed at this time in terms of bringing awareness to SCT. A key suggestion made during the meeting was that SCT educational efforts should avoid increasing stigma. It is essential that any educational initiative dedicated to SCT would not single out individuals who are trait carriers. An educational initiative would need to focus on the importance of raising awareness to SCT and the potential health implications associated with it. With this in mind, it is also important to be clear of the meaning of “tailored” education that was mentioned in the initial objectives of this study. By ultimately developing tailored education, our aim is to establish education based on the needs of our population in

order to increase generalized awareness to SCT. Tailored education it is not a means to single out individuals with SCT, rather it is a means to provide meaningful education that is developed with the input of community members statewide. In addition to avoiding stigma, a principal goal of SCT education should be to avoid worrying parents or individuals with SCT unnecessarily. It is essential to explain that the potential health implications associated with SCT are rare and preventable if proper precautions are taken.

In addition to the practice recommendations for moving forward with an educational program for SCT, there are many policy implications that can be derived from the results of this study. These policy implications range from the community level to the individual level such as issues related to testing and follow-up of people with SCT. A policy recommendation at the federal or national level may be to standardize educational information about sickle cell trait, particularly the information that is received during the newborn screening process. Currently each state produces its own educational materials about SCT to distribute to individuals and families, and the information provided within the education is not consistent among states. Some states will acknowledge that SCT can potentially have adverse health outcomes if proper precautions are not taken, and some states will simply express that SCT is clinically benign and that there is nothing to worry about. The inconsistency among educational materials among newborn screening programs could partially stem from the lack of medical consensus on the issue of labeling sickle cell trait as a potentially harmful condition. However, the medical literature clearly documents that there are rare, but potentially serious, health implications associated with SCT. Individuals who have SCT and their families must be made aware of these health implications during the newborn screening process. A key first step in accomplishing this would be to unify newborn screening educational materials to deliver a consistent message.

As the CDC meeting recommendations suggest, there needs to be an increased awareness of the issues surrounding SCT brought to the attention of policy makers. To adapt this message to policy

makers and other relevant stakeholders in the State of Nevada, it is important to put into context not only the general health implications surrounding SCT, and the lack of knowledge and awareness within our communities, but also the increasing prevalence of SCT and disease in Nevada. It may also be appropriate to discuss other inherent risks just from living in Nevada such as the climate and elevations in certain areas that may aggravate health issues associated with SCT. For example, the summer temperatures in Southern Nevada can be conducive to precipitating adverse effects related to SCT if someone is participating in strenuous activities, and some of the higher elevations in Northern Nevada may also be conducive to the potential adverse health outcomes related to SCT.

To bring light to these issues, a key recommendation that may bring increased awareness to policy makers within the State of Nevada would be to work with resources and actions that have already been established in our state, but are not being utilized; a primary action being to activate the Nevada Advisory Committee Concerning Sickle Cell Anemia (NRS.442.118). Implemented in 1989, this committee has been inactive since 1995, but one of the key activities of the committee as outlined in the Nevada Revised Statutes is to "Gather information regarding the need of screening and awareness programs and the operations of such programs" (Nevada Revised Statutes, 2013, para. 3). Furthermore, the committee is to, "make recommendations to the State Board of Education, State Board of Health, and others concerning the regulations necessary to implement these programs" (Nevada Revised Statutes, 2013, para. 4). The activation of the Committee Concerning Sickle Cell Anemia may be a major step in sickle cell awareness in the State of Nevada.

After increasing this awareness it is essential to address other pressing policy issues related to screening for SCT. Researching cost-effective methods where physicians, parents, or any individual could have access to their NBS results may make the idea of follow-up throughout the lifespan of the individual more feasible. In addition to increasing accessibility, a policy that can address the critical lack of genetic counselors and associated resources in our state is essential to move forward. Allocating

funds and support for these resources would help complete the critical link in the NBS follow-up cycle. Increased funding and support for the NBS follow-up process is essential because this would enable adequate and timely referrals for genetic counseling.

Implications for Future Research

There is limited research on community knowledge related to SCT. Future research should build on this study and the previous studies by conducting similar surveys with diverse populations to assess generalized community knowledge related to SCT. Our study focuses on parents of children with SCT and medical providers who care for children and their families affected with SCT. Since this study focused on a statewide sample, future studies wanting to focus on the State of Nevada, may want to examine specific regions within the State and understand if there is any variation among SCT resources and utilization, or population differences related to the familiarity of healthcare providers and treating SCT. Future research in the State of Nevada can also examine the knowledge of other variant traits, such as hemoglobin C trait, alpha thalassemia trait, as well as knowledge of sickle cell disease since the data regarding these conditions in the State of Nevada are available, but have yet to be reviewed. This would be also be important because educational information could be tailored based on the needs of the traits that individuals had concerns or lack of knowledge about.

Lastly, there needs to be an ongoing effort to implement educational programs regarding SCT and evaluating their effectiveness. Even though there is limited research and literature on community level SCT knowledge, the information is especially limited on the implementation of education programs and the effectiveness of these programs in relation to SCT.

Conclusions

There is a significant need for SCT education and resources in the State of Nevada. Provision of educational information can be accomplished in several ways, and this study has provided insight into the preferred methods of educational delivery and topics of interest from a sample of individuals who

are highly affected by SCT within the State of Nevada. As the results of this study have indicated, and in likeness to previous studies, a collaborative effort from all relevant stakeholders will be required to develop and implement innovative strategies that can educate communities and bring awareness to SCT in the State of Nevada.

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Appendix A

University of Nevada, Reno Institutional Review Board

Certificate of Approval

University of Nevada, Reno

Office of Human Research Protection

218 Ross Hall / 331, Reno, Nevada 89557

775.327.2368 / 775.327.2369 fax

www.unr.edu/research-integrity

Certification of Approval for New Protocol: Social Behavioral
Social Behavioral Institutional Review Board
FWA00002306

Date: July 22, 2013
To: Trudy Larson, MD
Copy: Yasmine Mohamednur

UNR Protocol Number: 2014S002
Protocol Title: Development of a statewide sickle cell trait educational campaign for parents and medical providers
Sponsor Names:

Type of Review: Expedited category 7 Minimal Risk

Meeting/Review Date: July 19, 2013
Approval Period: July 19, 2013 to July 18, 2013

This approval is for:
Approved number of subjects: 600
Approved documents: Protocol 070213, Provider survey invitation letter/information sheet 072213 (email and regular snail mail letter), Parent survey invitation letter/information sheet 072213, Key informant interview email, Provider survey, parent survey, key informant interview guide

The above-referenced protocol was reviewed and approved by one of UNR's Institutional Review Boards in accordance with the requirements of the Code of Federal Regulations on the Protection of Human Subjects (45 CFR 46 and 21 CFR 50 and 56).

Problems Researchers Must Report to the Research Integrity Office or IRB Staff
(to be reported as soon as possible, but within 10 business days)

- New or additional risks: Outcomes that the principal investigator believes are unexpected, related to the research, and suggest the research may place participants or others at greater risk of harm than was previously known or recognized
- Changes to expected harms or benefits: Any report indicating the frequency or magnitude of harms or benefits may be different than initially presented to the IRB
- Privacy: Any invasion of privacy related to an individual's participation in research
- Confidentiality: Any breach of confidentiality involving research data
- FDA Changes: Any change in FDA labeling or approval for a drug, device or biologic used in a research protocol
- Immediate harm: Any change to the protocol to eliminate an apparent immediate hazard to a research participant, prior to seeking IRB review and approval
- Prisoner: Any incarceration of a participant in a protocol not approved to enroll prisoners
- Sponsor: Any event that requires prompt reporting to the sponsor

- Sponsor: Any sponsor-imposed suspension for risk
- Protocol change: Any accidental or unintentional change to the IRB approved protocol that harmed participants or others, indicates participants or others may be at increased risk of harm, or has the potential to recur
- Device: Any unanticipated adverse device effect
- Department of Health: Any non-compliance identified by Department of Health audit or monitoring
- Federal agency: Any investigation or report by federal agency related to the research
- Medical license or practice changes: Any loss of license or hospital privileges by any researcher on the study
- Complaints: Any complaints that suggest participants or others may have been harmed or placed at increased risk of harm

PI Responsibilities

- Maintain an accurate and complete protocol file.
- Submit continuing projects for review and approval prior to the expiration date.
- Submit proposed changes for review and approval prior to initiation, except when necessary to eliminate apparent immediate hazards to subjects. Such exceptions must be reported to the IRB at once.
- Report any unanticipated problems which may increase risks to human subjects or unanticipated adverse events to the IRB within 5 days.
- Submit a closure request 10 days after project completion to the IRB.

Reference the protocol number on all related correspondence with the IRB. If you have any questions, please contact Valerie Smith at 775.327.2368.

For Veteran's Administration research only

VA Research: No
Flag VA Medical Record: N/A
NOTE: You are not approved to begin this research until you receive an approval letter from the VASNHCS Associate Chief of Staff for Research stating that your research has been approved by the Research and Development Committee.

Appendix B

Parent and Provider Surveys

Provider Survey

Introduction

You are invited to participate in a research study being conducted through the Nevada Division of Public and Behavioral Health's Newborn Screening Program. We would like to have you complete a short survey. The purpose of the survey is to assess the existing knowledge related to sickle cell trait of both parents and medical providers in the community. We would also like to get your ideas about how we can develop a statewide educational program addressing sickle cell trait.

We are contacting you because you were identified as licensed medical practitioner that interprets newborn screening results in the State of Nevada. We believe it would be beneficial to develop and educational initiative with your assistance to ensure you and all other medical providers throughout the state have the necessary education and support services to assist individuals and their families with sickle cell trait.

There are four parts to the survey. It should take you no longer than ten minutes to complete it.There are no risks with participating in this survey. You will not be asked to put your name or any other identifying information on this survey.

Please be assured that your individual survey responses will be kept safe and confidential.

Thank you for taking the time to complete this survey.

Part I : General Knowledge

Directions: Please select your answer for the following questions.

1. How would you rate your knowledge of genetics?

Poor ○ Fair ○ Good ○ Very Good ○ Excellent ○

General Knowledge Continued

2. Which of the following statements is true regarding sickle cell trait? (Please select all that apply).

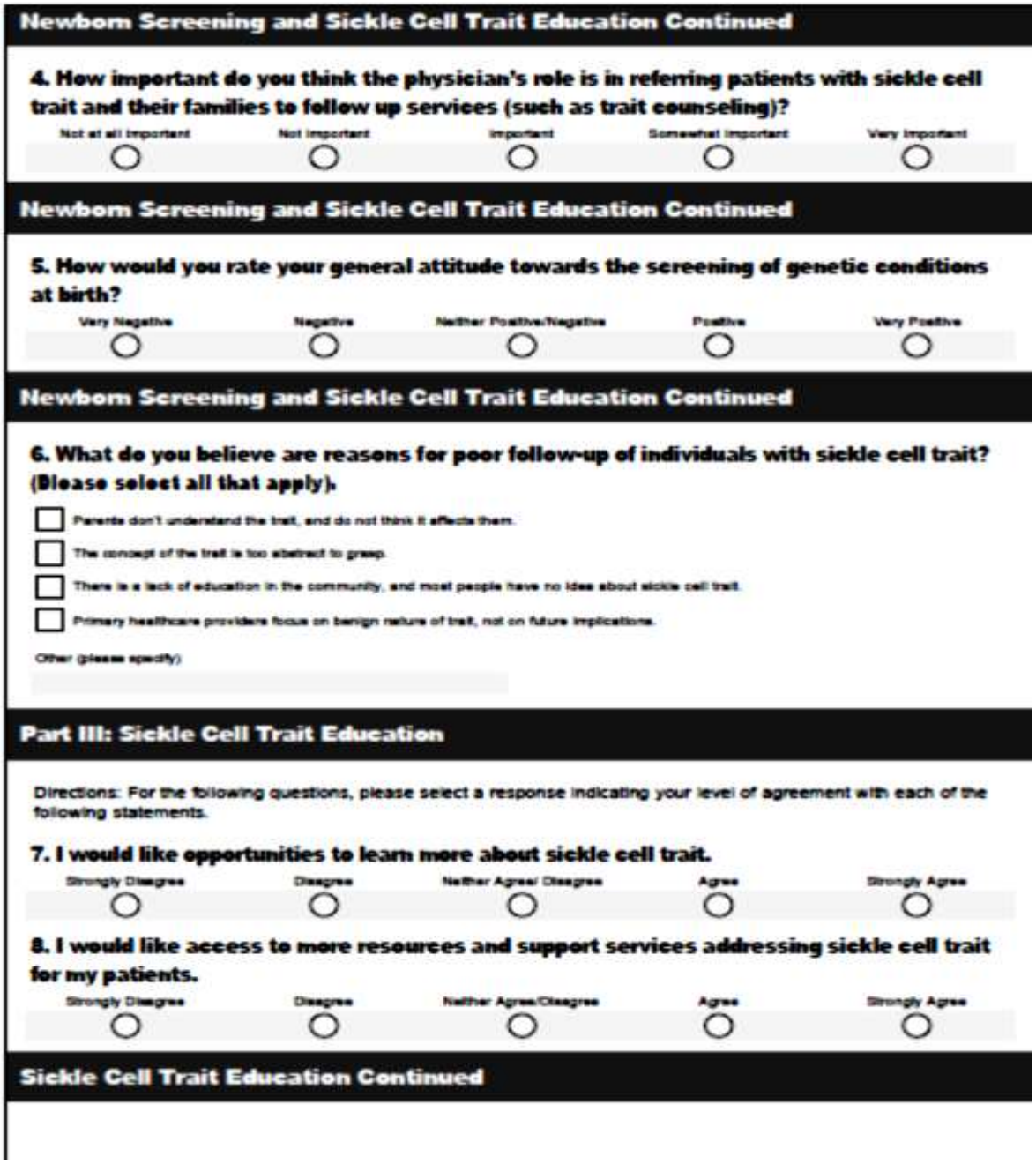
- ☐ Sickle cell trait is clinically benign and will never lead to any health problems.
- ☐ Sickle cell trait is a more severe form of sickle cell anemia.
- ☐ Only African Americans can inherit sickle cell trait.
- ☐ All of the above statements are true
- ☐ None of the above statements are true.

Part II: Newborn Screening and Sickle Cell Trait

Directions: For the following questions, please select a response indicating your level of agreement with each of the following statements.

3. How often do you see patients with sickle cell trait?

Not at all ○ Not Often ○ Often ○ Somewhat Often ○ Very Often ○



Newborn Screening and Sickle Cell Trait Education Continued

4. How important do you think the physician's role is in referring patients with sickle cell trait and their families to follow up services (such as trait counseling)?

Not at all important ○ Not important ○ Important ○ Somewhat important ○ Very important ○

Newborn Screening and Sickle Cell Trait Education Continued

5. How would you rate your general attitude towards the screening of genetic conditions at birth?

Very Negative ○ Negative ○ Neither Positive/Negative ○ Positive ○ Very Positive ○

Newborn Screening and Sickle Cell Trait Education Continued

6. What do you believe are reasons for poor follow-up of individuals with sickle cell trait? (Please select all that apply).

☐ Parents don't understand the trait, and do not think it affects them.

☐ The concept of the trait is too abstract to grasp.

☐ There is a lack of education in the community, and most people have no idea about sickle cell trait.

☐ Primary healthcare providers focus on benign nature of trait, not on future implications.

Other (please specify)

Part III: Sickle Cell Trait Education

Directions: For the following questions, please select a response indicating your level of agreement with each of the following statements.

7. I would like opportunities to learn more about sickle cell trait.

Strongly Disagree ○ Disagree ○ Neither Agree/ Disagree ○ Agree ○ Strongly Agree ○

8. I would like access to more resources and support services addressing sickle cell trait for my patients.

Strongly Disagree ○ Disagree ○ Neither Agree/Disagree ○ Agree ○ Strongly Agree ○

Sickle Cell Trait Education Continued

9. If an educational program were to be developed addressing sickle cell trait, what topics related to sickle cell trait would you be interested in?

	Little or No Interest	Some Interest	Strong Interest
Genetics of sickle cell trait	○	○	○
Follow-up services available	○	○	○
Trait counseling	○	○	○
Potential health problems associated with sickle cell trait	○	○	○
Preventive measures for potential health problems associated with sickle cell trait.	○	○	○

Sickle Cell Trait Education Continued

10. Please indicate the format/delivery that you have interest in for an educational program about sickle cell trait in the State of Nevada?

	Little or No Interest	Some Interest	Strong Interest
Mail out written information or pamphlet	○	○	○
Informational Community Meetings	○	○	○
Publicize on Television	○	○	○
Publicize on radio	○	○	○
Distribute a video or CD about Sickle Cell Trait	○	○	○
Website dedicated to Sickle Cell Trait education and resources	○	○	○
Community Based Events	○	○	○

Sickle Cell Trait Education Continued

11. Are there any other comments or concerns you have regarding a sickle cell trait educational program? Please type them below.

Part IV: Demographics

Directions: Please make the selection that best describes you. *Please Note: You are not obligated to answer these questions*

12. What is your current age?

- ◯ 18 to 24 years of age
- ◯ 25-34
- ◯ 35-44
- ◯ 45-54
- ◯ 55 or older

Demographics Continued

13. What is your gender?

- ◯ Female
- ◯ Male

14. Are you Hispanic or Latino?

- ◯ Yes
- ◯ No
- ◯ I'm not sure

15. What is your race? Please choose one or more.

- ☐ White
- ☐ Black or African-American
- ☐ Asian
- ☐ Native Hawaiian or other Pacific Islander
- ☐ American Indian or Alaska Native
- ☐ Other

Demographics Continued

16. How long have you lived in the State of Nevada?

- ◯ Less than 1 year
- ◯ 1-5 years
- ◯ Greater than 5 years

17. How long have you practiced medicine in the State of Nevada?

- ◯ Less than 1 year
- ◯ 1-5 years
- ◯ Greater than 5 years

Parent Survey

Introduction

You are invited to participate in a research study being conducted through the Nevada Division of Public and Behavioral Health's Newborn Screening Program. We would like to have you complete a short survey. The purpose of the survey is to find out how much parents and medical providers know about sickle cell trait. We would also like to get your ideas about how we can develop a statewide educational program about sickle cell trait.

We are contacting you because your child was identified as a sickle cell trait carrier during the years 2005-2012 in the State of Nevada through the newborn screening process. We believe that it would be helpful to have an educational campaign to help people learn more about sickle cell trait. We would like to have your help to do this.

There are four parts to the survey. It should take you no longer than ten minutes to complete it. There are no risks with participating in this survey. You will not be asked to put your name or any other identifying information on this survey. Please be assured that we will keep your survey responses safe and confidential.

Thank you for taking the time to complete this survey.

Part 1: General Knowledge

1. How would you rate your general knowledge of genetics?

Poor ○ Fair ○ Good ○ Very Good ○ Excellent ○

2. Have you ever heard of the term sickle cell trait?

○ Yes
○ No
○ I'm not sure

3. How do you think a person gets sickle cell trait?

○ It is contracted through the air (e.g. if someone sneezes, or coughs).
○ A person is born with it (it is hereditary).
○ A person gets it through a blood transfusion.
○ I am not sure how a person gets sickle cell trait.

4. Do you think the following statement is true or false?
"There is no difference between sickle cell trait and sickle cell disease."

- ◯ True
- ◯ False

5. Do you think the following statement is true or false?
"Sickle cell trait can turn in to sickle cell anemia."

- ◯ True
- ◯ False

6. Which of the following statements is true regarding sickle cell trait? (Please select all that apply).

- ☐ Sickle cell trait will never lead to any health problems.
- ☐ Sickle cell trait is a more severe form of sickle cell anemia.
- ☐ Only African Americans can inherit sickle cell trait.
- ☐ All of the above statements are true
- ☐ None of the above statements are true.

Part 2: Newborn Screening and Sickle Cell Trait

7. How does someone find out if they have sickle cell trait? (Please select all that apply).

- ☐ Through a blood test and screening process performed on newborn babies before they leave the hospital (e.g. heel stick test).
- ☐ If a person is older (ex. adolescent or adult), they can take a simple blood test if they want to know their sickle cell trait status.
- ☐ Through a swab of the mouth.

8. Do you have at least one doctor that you think of as your child's primary doctor or pediatrician?

- ◯ Yes
- ◯ No
- ◯ I'm not sure

9. Where do you get most of your health information. (Please select one).

- ◯ Medical provider (ex. a Doctor or Nurse)
- ◯ Internet
- ◯ Television
- ◯ Books/Magazines

Other (please specify)

_____

10. In the past five years have you received information about sickle cell trait from a medical source (ex. Doctor or Nurse)?

- ◯ Yes
- ◯ No
- ◯ I'm not sure

11. How would you rate your satisfaction with the information given to you regarding your child's sickle cell trait status by your health care providers?

Very Dissatisfied	Somewhat Dissatisfied	Neither Satisfied/Dissatisfied	Somewhat Satisfied	Very Satisfied
◯	◯	◯	◯	◯

12. Do you feel you have received adequate follow-up services (such as counseling, education, and support) for your child's sickle cell trait?

- ◯ Yes
- ◯ No
- ◯ I'm not sure

13. How would you rate your general attitude towards the screening of genetic conditions at birth?

Very Negative	Somewhat Negative	Neither Positive/Negative	Somewhat Positive	Very Positive
○	○	○	○	○

[redacted]

14. What is your level of agreement with the following statement?
"I would like opportunities to learn more about sickle cell trait"

Strongly Disagree	Disagree	Neither Agree/Disagree	Agree	Strongly Agree
○	○	○	○	○

15. What is your level of agreement with the following statement?
"I would like to have access to more resources and support services addressing sickle cell trait"

Strongly Disagree	Disagree	Neither Agree/Disagree	Agree	Strongly Agree
○	○	○	○	○

[redacted]

16. If an educational program were to be developed addressing sickle cell trait, what topics related to sickle cell trait would you be interested in?

	Little or No Interest	Some Interest	Strong Interest
Genetics of sickle cell trait	○	○	○
Follow-up services available for sickle cell trait	○	○	○
Trait counseling	○	○	○
Potential health problems associated with sickle cell trait	○	○	○
Measures to prevent for potential health problems associated with sickle cell trait.	○	○	○

17. Please indicate the format/delivery that you have interest in for an educational program about sickle cell trait.

	Little or No Interest	Some Interest	Strong Interest
Mail out written information or pamphlet	○	○	○
Informational Community Meetings	○	○	○
Publicize on Television	○	○	○
Publicize on radio	○	○	○
Distribute a video or CD about Sickle Cell Trait	○	○	○
Website dedicated to Sickle Cell Trait education and resources	○	○	○
Community Based Events	○	○	○

18. Are there any other comments or concerns you have regarding a sickle cell trait educational program? Please type them below.

Part 4: Information about You and Your Family

* You are not obligated to answer these questions*

19. How many children do you have?(Please type in the box below).

20. How many children do you have with sickle cell trait? (Please type in the box below).

21. Which category below includes your age?

- ◯ 17 or younger
- ◯ 18-20
- ◯ 21-29
- ◯ 30-39
- ◯ 40-49
- ◯ 50-59
- ◯ 60 or older

22. What is your gender?

- ◯ Female
- ◯ Male

23. Are you Hispanic or Latino?

- ◯ Yes
- ◯ No
- ◯ I'm not sure

24. What is your race? Mark one or more.

- ◯ White
- ◯ Black or African American
- ◯ Asian
- ◯ Native Hawaiian or Other Pacific Islander
- ◯ American Indian or Alaska Native
- ◯ Other

25. How long have you lived in the State of Nevada?

- ◯ Less than 1 year
- ◯ 1-5 years
- ◯ Greater than 5 years

Appendix C

Key Informant Interview Script

Key Informant Interview

Script

Introduction:

Thank you for meeting with me today,

My name is Yasmine Mohamednur. I am a student investigator working with the Nevada Division of Public and Behavioral Health's Newborn Screening program. I am working on a project where the aim is to bring more awareness and knowledge to potential health problems associated with sickle cell trait through the development of a statewide educational program.

An essential first step in our endeavor is to better understand the needs of the community, and in order to do this we are conducting interviews with individuals with whom we feel have a good understanding of the community needs. We believe that your knowledge of the community will be invaluable.

Please be assured that all of the information from this interview will be kept from inappropriate disclosure.

The interview should take between 45-60 minutes.

Interview Questions:

1. **Can you tell me about your background and your work in the field of genetics/ hematology/ newborn screening?**
2. **What major issues do you see in your clients with sickle cell disease or sickle cell trait in particular?**
    - Any major conditions/challenges faced by the target population (individuals with sickle cell trait)?
    - What has happened that makes you believe this is a problem?
        - Can you tell us more about this
    - Who does this problem/ problems affect?
        - How does it affect them? [Encourage specifics.]
    - What do you feel is the cause of these major problems?
        - How might education help?
    - Are there any other problems that come to mind?
    - Are there some themes that are evident in the health of the patients you treat?
    - Are there different issues for different segments of the population in Nevada?
    - Are there any language, or cultural issues that you feel need to be addressed?
3. **Can you describe any community advocacy or community involvement that is related to sickle cell trait?**
4. **What do you think can encourage support and more local community involvement around a health issue like sickle cell trait?**

5. **What do you think are barriers to community involvement surrounding a health issue like sickle cell trait?**
    - Complexity of the issue
    - Lack of information
6. **Do you think we have covered everything that you think is important? Do you have any recommendations of other people I should talk to?**

Closing:

Thank you very much for time, your knowledge will be very valuable and helpful for our program planning. Thank you again.

Appendix D

Open-Ended Responses

Table A. Reasons for Poor Follow-up of Sickle Cell Trait (Provider Survey)

Table B. Sources Where Most Health Information is Retrieved (Parent Survey)

Table C. Comments or Concerns Regarding Sickle Cell Trait Education (Provider Survey)

Table D. Comments or Concerns Regarding Sickle Cell Trait Education (Parent Survey)

[Provider Survey] What do you believe are reasons for poor follow-up of individuals with sickle cell trait? (N=3).

Question number six (provider survey) asked providers what they perceived as reasons for poor medical follow up of individuals with sickle cell trait. Participants were given a list of options they could select within the survey as well as an open-ended comment box. From four open-ended responses three were able to be categorized (N=3) and, two similar themes emerged, these were: minimal sickle cell trait awareness throughout the lifespan and medical costs. The following table displays the responses under each theme.

Table A. Reasons for Poor Follow-up of Sickle Cell Trait (N=3)

Minimal sickle cell trait awareness throughout the lifespan	Medical costs
It is diagnosed at birth and most patients are asymptomatic and people think they can wait until marriage/children before they need to deal with it.	Lack of medical insurance and cost of healthcare.
Primary healthcare providers and parents do not discuss sickle cell trait as the child grows older, and probably the awareness fades in memory of the parents and the providers who have the trait.	

[Parent Survey] Where do you get most of your health information? (N=8)

Question number nine (parent survey) asked parents what sources they received most of their health information from. Participants were given a list of options in the survey as well as an open-ended comment box to enter additional sources that were not listed in the survey. From eight open-ended responses, eight responses were categorized (N=8). Three themes emerged, these were: website/social media, literature, and other (could not be grouped). The following table displays the responses under each theme. Some of the comments were grouped under more than one theme.

Table B. Sources Where Most Health Information is Retrieved (N=8)

Website/ social media	Literature	Other
Internet	Books and Magazines	From husband who has personal knowledge
Internet	Books and Magazines	Television
Internet	Books and Magazines	Children's Hospital of Oakland, Ca.
	Reading	College/University

[Provider Survey] Are there any comments or concerns you have regarding a sickle cell trait education program? (N=4).

Question number eleven (provider survey) asked providers if they had any comments or concerns regarding a sickle cell trait education program. Of the six open-ended responses, four (N=4) were able to be categorized, and three themes emerged. These themes were: cost concerns, suggestions for educational resources, and clinical issues related to sickle cell trait. The following table displays the responses under each theme.

Table C. Comments or Concerns Regarding Sickle Cell Trait Education (N=4)

Cost concerns	Suggestions for educational resources	Clinical issues related to sickle cell trait
Is this a free service? Is it covered by insurance?	Need an easy to read and understand website that parents can use and that we can print handouts from. Maybe have names and addresses and phone numbers of genetic counselors and doctors they can follow up with	Clinicians need to know that every individual should be aware of their status as everyone is screened at birth.
	There has to a concerted effort to education the community with physicians, families, and form collaborations with concerned groups.	

[Provider Survey] Are there any comments or concerns you have regarding a sickle cell trait education program? (N=6).

Question number eighteen (parent survey) asked parents if they had any comments or concerns regarding a sickle cell trait education program. Of the eight open-ended responses, six (N=6) were able to be categorized, and two themes emerged. These themes were: uncertainty regarding personal trait status, and need for more educational resources. The following table displays the responses under each theme.

Table D. Comments or Concerns Regarding Sickle Cell Trait Education (N=6)

Uncertain of personal trait status	Need for more educational resources
I would like to know what the cut-off number is since I was told I was on the border line, and I need to know if I have the trait or the disease.	My husband has sickle cell trait; we knew that by having a child our child would inherit the trait. So far our son has not had any problems, but I am interested in more information to help as he grows to understand the trait that he carries.
I have a mutation of the trait which does affect my iron levels. It is because of this that I knew more about the trait but it was only diagnosed later in life before that I was incorrectly diagnosed with anemia	I think that brochures provided by pediatricians or hospitals after the PKU test would be effective. Keep information at the 8th grade level since the average American has less than mediocre education level and subsequent intelligence. -
	I would like to see an educational program encompassing both sickle cell trait and sickle cell anemia; it is very disappointing that doctors in this area (Northern Nevada), even hematologists have little to no practical experience or knowledge of sickle cell in any form. The more information the better!! -
	There should be more information from the hospital when leaving with a newborn with sickle cell trait, especially if you cannot afford more help

Appendix E

PRECEDE-PROCEED Framework Sickle Cell Trait Education

Primary Research Question	**What are the preferred educational approaches of parents and medical providers that can be used to develop tailored sickle cell trait education that will generate and enhance awareness of the potential health risks associated with sickle cell trait throughout the State of Nevada?**

PRECEDE/PROCEED FRAMEWORK **PRECEDE- PHASES 1-4**		
Phase	**Research Questions**	**Activity**
1: Social Assessment	1. What is the current level of community awareness of the risks associated with sickle cell trait? 2. What are parents/providers knowledge and perceptions of sickle cell trait screening and counseling. 3. What concerns do parents/providers have regarding sickle cell trait screening? 4. What do parents of children born with SCT need in regards to sickle cell trait education? 5. What do medical providers who care for children with SCT need in regards to sickle cell trait education? 6. Are individuals with sickle cell trait receiving adequate follow-up services in the state of Nevada?	1. Surveys Parents of Children with SCT (State of Nevada) 2. Survey of Providers who interpret newborn screening results for sickle cell trait in the state of Nevada. 3. Key Informant Interviews
2. Epidemiological Assessment	1. What are the genetic, behavioral, and environmental factors that may influence individuals with sickle cell trait? 2. How will identified (genetic, behavioral, and environmental) factors influence the knowledge and perception of sickle cell trait among parents/ medical providers?	1. Databases 2. Literature Review 3. Data and Statistics from .gov websites and other credible/ peer reviewed sources

Phase	Research Questions	Activity
3. Educational & Ecological Assessment	1. What are the predisposing, reinforcing, and enabling factors that influence medical providers/ parent's knowledge and perceptions of sickle cell screening and counseling?	1. Categorize and select the predisposing, enabling, and reinforcing factors that seem to have a direct impact, and greatest potential to change the behavioral and environmental targets generated from previous stages.
4. Administrative & Policy Assessment and Intervention Alignment	1. What resources exist an what resources will be required to develop an educational campaign that can increase medical provider and parental awareness of the risks associated sickle cell trait in the State of Nevada?	1. Policies 2. Existing and needed resources 3. Organizational barriers/limitations to overcome

PROCEED- PHASES 5-8

Phase	Research Questions	Activity
5. Implementation	1. Develop educational program based on survey data	1. Develop and Implement educational Program.
6. Process Evaluation	1. Continued as program being implemented	1. Assess participant satisfaction of program 2. Identify potential barriers to success.
7. Impact Evaluation	1. Assess any immediate changes that can be attributed to the educational program (i.e. increase in knowledge regarding SCT, follow-up surveys).	1. Assess immediate effects of program on target behaviors and their predisposing, reinforcing, and enabling factors, and the impact on environmental factors.
8. Outcome Evaluation	1. Long term effects of the educational intervention.	1. Assess quality of life and epidemiological indicators identified at the beginning of the process (morbidity, mortality, and disability).