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Reproductive decisions in people with sickle cell disease or sickle cell trait

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Abstract

In the context of an inherited condition such as sickle cell disease (SCD), it is critical to understand how people with SCD or carriers (sickle cell trait [SCT]) face the challenges of making informed reproductive health decisions. The purpose of this analysis was to examine the beliefs, attitudes, and personal feelings of people with sickle cell disease or sickle cell trait related to making informed reproductive health decisions. We conducted three focus groups with a total of 15 people who had either SCD or SCT. We identified five themes: health related issues in sickle cell disease; testing for sickle cell trait; partner choice; sharing sickle cell status with partners; and reproductive options. These findings enhance understanding of the reproductive experiences in people with SCD and SCT and provide the groundwork for developing an educational intervention focused on making informed decisions about becoming a parent.

In the context of an inherited condition such as sickle cell disease (SCD), it is critical to understand how people with SCD or carriers (sickle cell trait [SCT]) face the challenges of making informed reproductive health decisions. Currently, more than 70,000 Americans are diagnosed with SCD and an additional 2.5 million African Americans are living with SCT (Sickle Cell Disease Association of American, Inc. [SCDAA], n.d.). SCD consists of a series of closely related inherited hemoglobin disorders including homozygous SCD (SCD-SS;

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HbSS), sickle hemoglobin C disease (SCD-SC; HbSC) and two common types of sickle beta thalassemia (SCD-S β^0 -thal, HbS β^0 -thal; SCD- β^+ -thal, HbS β^+ -thal; see Table 1). SCD and SCT are most commonly found in people of African, Mediterranean, Middle Eastern and Indian (sub-continent) origin. In the blood circulation of affected individuals, as red blood cells release their oxygen to the tissues, the deoxygenated sickle hemoglobin molecules begin to aggregate, literally freezing the cells into stiff, sticky crescent-like shapes resulting in blockage of blood vessels and severe pain. Other problems can develop such as chronic anemia, organ damage, infection, stroke, acute chest syndrome, and leg ulcers. No universal cure for SCD has been developed, but treatment for symptoms and complications is available.

Parents transmit SCD and SCT to their children in an autosomal recessive manner (see Figure 1). In the common form of the disease, the sickle cell gene ($\beta_6^{\text{glu} \rightarrow \text{val}}$) encodes an abnormal hemoglobin (hemoglobin S) and is inherited from both biological parents. In other SCD variants, the child inherits a sickle hemoglobin gene from one parent, and another abnormal hemoglobin gene from the other parent (see Table 1 for examples). People who have SCT do not have SCD and usually do not have symptoms because they inherit one normal gene from one of their parents. Hemoglobin electrophoresis, isoelectric focusing and high performance liquid chromatography (Pack-Mabien & Haynes, 2009; SCDA, n.d.), and used less commonly, DNA-based testing (Wethers, 2000) can determine a person's sickle cell status.

In the United States, all states and the District of Columbia test infants for SCD through newborn screening (NBS) programs (genes-r-us.uthscsa.edu). Because young children with SCD are susceptible to life threatening infections, NBS for SCD is intended to identify infants who would benefit from early treatment with penicillin and close monitoring of symptoms to prevent complications (Pass et al., 2000). But screening for SCD also identifies infants who have SCT, who are generally asymptomatic and may not benefit early in life from knowing their sickle cell status (Miller et al., 2009; Pass et al.), especially before they become sexually active. Parents, too, may benefit from knowing the results of screening for SCT and being offered relevant information, education and counseling to increase their understanding of SCD inheritance as they consider future reproductive decisions.

With improved diagnosis and health care, many more children with SCD now grow into adulthood, where they, as well as people with the trait, face serious decisions surrounding childbearing. Pregnancies might occur with little forethought or opportunity to make well-informed reproductive health decisions which, as found in previous studies (Acharya, Lang & Ross, 2009; Gallo et al., 2008), are likely related to insufficient or incorrect knowledge about the transmission of inherited conditions including SCD or SCT.

Reproductive health decisions and behaviors are complex and have been studied extensively in adolescents and adults (Ahluwalia, Johnson, Rogers, & Melvin, 1999; Joyce, Kaestner, & Korenman, 2002). However, there are few studies published to date about people with SCD or SCT and their intentions to become a parent (Asgharian & Anie, 2003). People with the SCD or SCT have the options to not have a child, to have a child with or without SCD or SCT, or to seek other non-childbearing options such as parenting an unaffected, non-biological child (e.g., foster, adopt). The decision to have children is influenced by people's knowledge of the genetic transmission of SCD or SCT and their perceptions of the disease severity and risks of pregnancy to the mother, as well as the people's attitudes and beliefs about preserving the family genetic heritage (Asgharian & Anie). An approach to encourage informed reproductive health decisions by people with SCD or SCT is to enhance their reproductive health knowledge with tailored information so that their reproductive health behaviors are consistent with their reproductive health intentions.

We are conducting a study of adults with SCD or SCT using a new web-based, tailored, multimedia education program about the disease and trait, health consequences, and reproductive options (citation blinded for review). The educational program is designed to help men and women with SCD or SCT implement a parenting plan that will support their informed reproductive health decisions and reproductive health behaviors.

To prepare for the larger study, focus groups were conducted with adult women and men with SCD or SCT. Whereas the intent of the focus groups was to ensure that the questionnaires and the educational intervention would be sensitive, effective, relevant, clear, and culturally appropriate for the intended population, the focus groups also elicited beliefs, attitudes, and personal feelings surrounding SCD or SCT and reproductive decisions. Therefore, the purpose of this analysis was to examine the beliefs, attitudes and personal feelings of people with SCD or SCT related to making informed reproductive health decisions.

Methods

Sample

Using purposive sampling, participants were recruited from patients with SCD attending an adult hematology specialty clinic for routine treatment, parents of children with SCD who attended a pediatric hematology specialty clinic for routine treatment, and people from the community who had SCT. All participants were recruited in the greater metropolitan area of a large, urban Midwest City by direct contact where a provider introduced the patient or person with SCT to the research team. Eligible participants were 36 years of age or older, and able to speak and read English. People who were 36 years and older were selected because they were past the main childbearing years and were expected to have more informed ideas on the general aspects of the study questionnaires and intervention.

The composition of the focus groups was directed toward sickle cell status with participants with SCD, parents of children with SCD, and people with SCT from the community composing separate focus groups. The participants were separated into groups so that answers of people with specific experiences would not affect the answers of others with different experiences. No demographic information was collected from the participants on the number of children in their family or their conscious effort to have or not have children.

Data Collection

Prior to data collection, the institutional review board at the investigators' university approved the study. Focus groups were held at the medical facility where participants were familiar with the setting. After consenting in writing to participate in the study, participants completed a short demographic questionnaire prior to each group session. Questionnaire items (see Table 2 for examples) were presented and the general content areas of the educational program to the group using Power Point slides. Each participant also had a handout of the slides.

The study team included an experienced group facilitator who was African American. The group facilitator followed each slide with the following questions: What do you think about when you hear or read this question? Should we ask this question? What are the suggestions for changing the wording? Only three focus groups were conducted because the participants' responses became repetitive and offered no new information. Each focus group session took 2–3 hours and each participant received \$50 at the conclusion of the session.

Analysis

The analysis in this article addresses the beliefs, attitudes, and personal feelings of adults who were 36 years of age or older with SCD or SCT as they responded to the study questionnaire items only. The audio recorded focus group interviews were transcribed verbatim and then the transcripts were processed and coded using the procedures of thematic analysis. Field notes recorded by members of the study team who attended the focus groups were used as a supplemental source of data. A graduate student checked the transcripts for accuracy by verifying them with the taped interviews. Two team members identified preliminary topical content and areas related to the participants' beliefs, attitudes, and personal feelings based on their interpretation of the study questionnaire items. Data analysis included reading each interview transcript in its entirety for initial descriptions of the content in each of the focus groups. A series of matrices were developed that summarized content by focus group and topical area and included representative statements to compare within and across the focus group responses (Miles & Huberman, 1994). Because the answers were very similar across all three focus groups, the themes and subthemes were developed from the process of reviewing, synthesizing and clustering all the participants' responses together from all three groups. The topical themes, subthemes and matrix entries were audited by the first two authors and all authors reviewed the final themes and reached consensus that they accurately reflected the data.

Results

Sample

The sample consisted of a total of 15 people with SCD ($n = 5$; 33%) or SCT ($n = 10$; 67%) who participated in three separate focus groups ($n = 5$; $n = 2$; $n = 8$). The majority of participants were female ($n = 11$; 73%) and African American ($n = 14$; 93%) with one Hispanic participant. Ages of the participants ranged from 36 to 63 years with a mean age of 47.7 years. The five people with SCD had varying types of SCD: homozygous sickle cell disease (SCD-SS, HbSS) ($n = 2$); sickle hemoglobin C disease (SCD-SC, HbSC) ($n = 2$) and sickle beta plus thalassemia disease (SCD- β^+ -thal, HbS⁺ thal) ($n = 1$). Most of the participants with SCT had a relative or friend with SCD ($n = 9/10$).

Descriptive Themes

Five descriptive themes emerged from the focus group interviews: health related issues in sickle cell disease; testing for sickle cell trait; partner choice; sharing sickle cell status with partners; and reproductive options.

Health related issues in sickle cell disease—Despite recent improvements in treatments for SCD, many people in each focus group talked about the staggering health effects and complications of SCD in themselves, their children, or others. Health effects primarily addressed the “suffering” and “struggle” of pain episodes and complications accompanied by frequent visits to the emergency department and repeated hospitalizations; some participants noted that women with sickle cell disease can have difficult pregnancies. A few participants revealed that family members had died from SCD at an early age. As one participant with SCT remarked, “Most people don’t know how horrible [sickle cell disease] is, or can be ...” Within this context, participants did not want their children and grandchildren enduring the effects of SCD with some participants being “grateful” that their children did not inherit it. Participants noted that treatment is available for those who have SCD and that current treatment is much better now than when they were younger.

The participants had a range of emotions related to having the disease or trait. Some participants noted that they blamed themselves or indicated that their parents blamed

themselves for transmitting the disease to their children. As one participant with SCD shared,

My mother had ten of us, two with the disease and one has the trait. My brother is dead now. And every time I go into crisis my mother blames herself. Every time I get sick, she blames herself. So as much as I know she loves me, she knows I am going to suffer ... And I don't blame her and I tell her that all the time. She seems to never be able to forgive herself.

A few participants noted that they feared death from the disease and other people used "denial" to get through bad times. Knowing that caring for a child with the disease would be a "struggle," others accepted the disease as part of one's destiny. As one participant with SCD indicated,

You have to get to a point that you accept that it is God's will, and that He's not going to bring you to something He is not going to bring you through.... This is not the only struggle and life goes on. And it gets better; it is an up and down life.

Most participants expressed a major concern about young people's lack of understanding about SCD and SCT. They consistently embraced educating young people early as the key to prevent genetic transmission of SCD; broadening young understanding of the differences between the disease and the trait; and teaching basic information, including SCD is not contagious like a sexually transmitted infection. As one participant with SCT noted, "And some [people say], 'what's that? I didn't catch that from nobody. If he has it, I am going to stay away from [him],' and I say, 'You can't get it like that' ..." Several participants thought that even though parents might raise objections, young children, 8 and 9 years old, need sickle cell education because many are sexually active at a "very tender age." As one participant with SCT explained,

Sometimes you have to hit hard about this. You're not saying it is just how it is. You can't be afraid of somebody's feelings if their mother and father are going to think their child has no business learning about this because she is only 8 years old ... we need to get right to the point and right to the meaning. This is what you need to know and this is why you need to know it.

A few participants indicated that, in general, people perceived SCD and SCT as "just a minority" disease and they needed to be aware of the possibility of having SCT. One participant with SCT thought that there were "not...enough heroes" or "role models" with SCD or SCT for the public to recognize that "sickle cell is a major concern for America, not just minorities." A Hispanic woman with SCT who had a child with SCD was quite vocal about other ethnicities and races learning about the risk for having the trait because she did not know she carried the trait before she became pregnant with her son who had the disease. She said, "Education needs to be spread not only to African Americans but to Latinos, Hispanics, and Indians ... because they are susceptible to get the trait or the disease [too]. ..." She also indicated that she was told by her health care provider that SCD was only an "African American disease" and when she found out that her newborn baby had SCD, she was "devastated" and would have considered terminating the pregnancy.

Testing for sickle cell trait—Most participants agreed that getting tested for SCT before pregnancy can give individuals and couples important information about reproductive options rather than "taking the chance" of having a child with the disease. One participant with SCD indicated, "If you are not sure if it runs in your family, go get tested." Along this same line, another participant with SCD addressed her own personal situation related to her partner's uncertainty about his sickle cell status and getting tested, "And when I asked him,

he said, 'I don't have that.' and I said, 'How do you know you don't have that?' And then when they tested him, he found out he had it."

Participants pointed out several major reasons why people do not get tested for SCT, including lack of understanding about the effects and inheritance of SCD and SCT in the family and fear of needle sticks. As one participant with SCT noted, "A lot of people need to know [their sickle cell status] if they do this baby thing, but they are scared to be stuck with the needle.... even when [they] have all these tattoos!" They also thought that young people did not appreciate the importance of knowing their sickle cell status and the reproductive options they might have available.

Partner choice—Within the context of learning early about their sickle cell status and sickle cell inheritance, participants understood and agreed that choosing a partner without disease or trait prevents children from being born with the disease. One of the male participants with SCD revealed, "I just want to say, sickle cell disease [can be prevented] because of the choice we have once you found out you have the trait. I found out early on, so I had to look for women who didn't have the trait." Another participant with SCT indicated that having the trait is not necessarily a problem but choosing a partner without disease or trait is a core decision, "I would say that having trait is not a bad thing necessarily as long as you are informed and educated and you're careful about your [partner] choices."

Participants continued to voice concerns about young people lacking the knowledge and understanding about sickle cell inheritance, and their lack of awareness and concern about partner choice. For example, one participant with SCT said, "Young people don't talk about [their sickle cell status to others]; they just want to have fun. Kids don't care about a lot of things like sickle cell safe sex."

Sharing sickle cell status with partners—To facilitate partner choice, participants stressed the importance of talking about and sharing their sickle cell status with partners when their relationship got "serious" and/or "intimate," and when they wanted to begin a family but before pregnancy. They emphasized that sharing was a two-way process and that asking partners about their sickle cell status was quite appropriate. As one woman with SCT said, "You need to ask them if they have the trait because they are susceptible to having the trait or the disease." Most participants believed that even though they may have been reticent to share their sickle cell status early in the relationship it avoided surprises that might frighten their partners, such as frequent, unexpected sickle cell pain episodes. A woman with SCD noted,

I was skeptical at first telling my future husband, but I know that I have frequent crises and it would freak him out. When I told him, he said, "So what's going to happen?" and I said, "Some days you might see me dragging or I might be in a lot of pain." And he said, "What can I do?" And I said, "Just be here to comfort me, if I need for you to rub alcohol on my legs or arms. He was terrified because he said he never dated anyone with sickle cell [disease]. So when I did have my first crisis with him, and it was a bad one, he was hysterical. [I said to him], "It was nothing that you did; it's just something that happens".

Sharing early in the relationship was seen as contributing to discussions about continuing the couple's relationship, having children and available reproductive options. One participant with SCD said,

If I see a relationship that I think maybe I want to stay in, I just automatically say, "Look, I have sickle cell [disease]," so I tell them right off the bat. I do that because

I know that if you tell them right off the bat then at least they [can decide if they want to have] a relationship with [me]. I just want them to know my situation.

However, participants perceived that a person's age influenced the ease or difficulty of talking about or asking about a partner's sickle cell status. Most participants remarked that now that they were older and more mature it was easier to share this information with others. But based on their past experiences they sensed that for young people, the fear of rejection or being teased by others and the potential of being treated differently might hinder their ability to share their sickle cell status with their partners. For example, one participant with SCD remarked, "I wouldn't tell anyone because I used to hate getting treated differently. I wanted to be treated normal. That was the reason why I didn't tell...." Grandparents thought that it was easier for them to talk with their teen grandchildren about their own sickle cell status and the disease itself. They indicated that sharing this information can help teen relatives talk with others about their status. One grandparent who had a granddaughter with SCD noted,

Right now, my granddaughter is 15, and she is starting to accept it a little more. Like, "this is what she has." She will tell her friends now that she has sickle cell disease where before she tried to hide it. I told her I had sickle cell disease and she has a little knowledge about it now, so, it might be easier for her to talk about it.

One participant with SCD was so resolute that her children with trait talked to their partners that she took it on herself to share this information,

I made sure my children talked with whoever they were dating and let them know [they had the trait]. Even if my kids didn't tell them, I would ask the person and I would literally tell their parents. I did it because I don't want my grandkids to suffer like I suffered.

Reproductive options—Women participants expressed a strong desire for having biological children even when a clear risk of having a child with the disease was present. As one woman with SCD emphatically stated, "It's very rare that a woman doesn't want at least one child.... I would have to take the chance of bringing the child into this world with the disease." Another woman with SCD who wanted a child entrusted the outcome to God, "I have one daughter, praise God. I love her dearly and she has the trait. I dare to say that's how badly I wanted children. I wanted a child and God gave me one." Some participants revealed that they might have made different decisions about having children now knowing how severe the symptoms and complications of the disease could be. One woman with SCT, whose husband did not know he had the trait before having children explained,

If I had known that me and my husband had trait, we could have sat down and discussed it. We could have said, "Let's not bring another child into this equation." I could have made some educated decisions to not have children; maybe we could adopt or use other methods.

Whereas some women clearly expressed the desire to have children, some participants decided not to have children because of their perceived severity of the disease. One male participant with SCT who had made a decision early in his life not to have any children, even with the trait said, "I didn't have any kids and that was one of my decisions. By seeing first hand what the trait did to my sister, [I decided not to] have kids with the trait."

For most participants, having children with SCT was acceptable because they did not exhibit the signs and symptoms of the disease. But a couple of participants would not even take the risk of passing the trait to their children, for example, one participant with SCT said, "I will not risk even the trait ... if I knew my partner had the trait and I had the trait and there is a

possibility that my child could have [the trait], I will not risk putting my child through that experience.”

Some participants believed that having prenatal testing to determine if a fetus had the disease was a good idea, especially to prepare “psychologically for what’s to come.” However, the participants who had been offered an amniocentesis to identify the disease in their fetus declined the test. They turned down the test primarily because of their desire to have a baby and they feared that the fetus would be harmed. As one woman with SCD perceived,

I think it would be hard on a pregnant mother, especially a mother that always wanted a baby.... Most women like that are likely not to do anything until the baby comes out. They would be afraid that the test would poke in the wrong way and hurt the baby.

But participants indicated that asking the father to get tested prior to the birth was an alternative way to identify the possibility that the fetus had the disease; others decided to wait until the baby was born for newborn testing hemoglobin results.

Deciding to terminate a pregnancy because of the disease in the fetus or when the mother’s health is compromised by the disease was viewed as a difficult decision for most of the participants, but several were willing to consider the possibility especially in “life or death” situations. Major reasons for not terminating a pregnancy included religious beliefs that did not allow termination, and personal beliefs, such as “I don’t believe in abortion.” Whereas one participant pointed out that most people did not decide to terminate until they are placed in the situation, another participant replied that termination is a private matter and some people terminate pregnancies even if religious or personal beliefs do not allow it. One woman with SCD who shared with the group that she had an abortion lamented on her experience and the difficult decision she made 18 years ago,

With me it is hard, because I experienced [an abortion]. It wasn’t so much because of my parents; it was what the doctors were telling me. At the time I was just scared. So I didn’t want anything to happen to me and I didn’t want anything to happen to my baby. But when it was done it was always on my mind. I don’t believe in abortion but at the time I was terrified. I didn’t want a child to come into the world to suffer.

Participants had a range of knowledge and acceptability of reproductive options with some discussion centering on the use of temporary birth control methods versus more permanent method such as tubal ligation. Several of the women already had a tubal ligation after having a child and others were considering it. All the participants were unaware of *in vitro* fertilization (IVF) with preimplantation genetic diagnosis (PGD) as a reproductive option. They had many questions about why and how these procedures were done, the pain involved and costs. Whereas some participants did not see IVF with PGD as a possible option for themselves, a few considered it a possibility for others, as one woman with SCT said, “It might not be my choice, but it might be the next person’s.” For the most part, participants believed that adoption and foster care was usually not an option for young people because of their desire to have their own biological children.

Discussion

In general, most previous psychosocial research on SCD and SCT has been focused on the effects of the disease on people or issues of disclosing newborn screening results. The findings of our study illustrate that older women and men with SCD or SCT have diverse beliefs, attitudes and feelings about SCD and SCT and reproductive decisions when

responding to questionnaire items. As in the few other studies about reproductive decision making by people with SCD and SCT (Ahmed, Atkin, Hewison, & Green, 2006; Atkin, Ahmed, Hewison, & Green, 2008; Gustafson, Gettig, Watt-Morse, & Krishnamurti, 2007), our participants based their reproductive and testing decisions within the context of their previous experience with the condition, and their perceived severity of SCD that causes suffering from frequent pain episodes, harmful health effects and disease complications. However, for some participants, decisions were more complicated by the desire to have children and the more recent availability of treatments that provide improved quality of life and longevity (Atkins et al.).

For some of our participants, religious beliefs, norms and practices were important influences in deciding to have prenatal testing and/or terminating a pregnancy. For others, it was not predefined or fixed by religious beliefs. These findings are similar to those of other investigators who note that individual preferences, personal moral judgment, and beliefs and values took precedence over religious beliefs (Ahmed et al., 2006; Atkin et al., 2008). Our finding that a barrier to partner testing for SCT was the perception of pain associated with the phlebotomy and difficulty in convincing their partners to get tested was similar to findings by Gustafson and colleagues (2007).

Whether people with SCD or SCT had a strong desire to have children or not, most women participants understood the conflict between wanting a biological child and risking the chance of having a child with the condition. Similar to findings by Hill (2003) and Asgharian and Anie (2003), women in our study placed a high value on motherhood and their ability to have children, which allowed them to build their own families and family ties.

Although our participants considered knowing and sharing sickle cell status as important to partner choice before pregnancy, they indicated that in reality it is difficult for young people to make these decisions. These findings are similar to Asgharian and Anie (2003), and Duffy (n.d.) who found that both men and women with SCT had great difficulty discussing their status with partners because of the potential stigma associated with having SCT (Acharya et al., 2009) and fears about premature assumptions that the relationship was serious.

In contrast to our findings that knowing trait status was perceived as beneficial for planning pregnancy, African American women attending an obstetrics and gynecology clinic reported a general benefit of screening for sickle cell trait but did not see themselves, their partners or their children as susceptible to the disease (Gustafson et al., 2007). They also found a positive relationship between an understanding of recessive inheritance and perceived susceptibility to sickle cell disease. Unlike other studies, women and men in our study had a keen interest in preventing the transmission of the disease through educating young people early to know their sickle cell status before a pregnancy, to share their sickle cell status with partners and to consider choosing a partner without the disease or trait to avoid having a child with sickle cell disease.

In our study, the older adults with SCD or SCT also perceived their younger counterparts as being unconcerned about the effects of SCD and needing more education about its inheritance. As reported in a recent Pew Research Center telephone survey, almost 80% of those surveyed currently believe that the “generation gap” has widened between the point of views of older and younger people about social and moral values (Taylor, Morin, Parker, Cohn, & Wang, 2009). The insights of the older adults about the potential for grandparents to effectively reach African American teens and young adults has profound implications for interventions on planning for parenthood.

Together these findings are suggestive that education for young people about testing should also include information regarding the recessive nature of sickle cell inheritance and the relatively high prevalence of SCT in people with African, Hispanic, Mediterranean, and Indian origins. Furthermore, health care professional education is still needed about these same ideas.

Several study limitations warrant consideration. Although the focus group participants were recruited from a variety of settings and reached data saturation, the study results might not reflect the experiences of those who are uncomfortable sharing information in a group research setting. Also our study did not include people without personal experience of SCD and who might not understand the inheritance of SCD or SCT. Study results might also reflect the previous education that participants could have received about SCD and SCT, which might have influenced their acceptance of testing. Because the focus groups were conducted for the purpose of getting opinions and input on closed-ended questionnaire items and an educational program for a larger study, participants might not have fully described their experiences about their reproductive decision making and reproductive options about becoming a parent because the format may not have elicited that dialogue as readily.

In summary, we present evidence from older women and men with SCD or SCT of their diverse beliefs, attitudes and feelings about reproductive decisions. They expressed strong opinions about the need to reduce transmission of the disease by educating young people of their sickle cell status before a pregnancy, to share their sickle cell status with partners and to consider choosing a partner with normal hemoglobin to reduce the burden of sickle cell disease on their children. Our research team will use these findings to develop and test an educational intervention about reproductive options available to men and women who themselves have inherited either SCD or SCT.

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	A	S
A	AA	AS
S or X	AS or AX	SS or SX

Figure 1.

Example of a Punnett Square probability for a couple at risk for a child with SCD and other hemoglobin variants (X). If both parents have SCT (AS), or if one parent has SCT and the other parent has another hemoglobinopathy trait (AX), then with each pregnancy, there is a 25 percent chance (1 in 4) that the child will have SCD (SS) or a SCD variant (SX), 25 percent chance (1 in 4) that the child will have normal hemoglobin (AA), and 50% chance (1 in 2) that the child will be a hemoglobinopathy carrier (AS or AX). A = normal hemoglobin; S = sickle cell hemoglobin, X = hemoglobin variant (e.g., C, β^0 -thal, β^+ -thal). Adapted from <http://www.ctbiobus.org/TestSite/BioBus/download/punnet.pdf>

Table 1

Types of Sickle Cell Disease

Condition	Condition acronyms	Inheritance/Clinical manifestations
Homozygous sickle cell disease, sickle cell disease SS, or sickle cell anemia	HbSS or SCD-SS	Hemoglobin S gene is inherited from both parents. Usually the most severe form of sickle cell disease.
Sickle hemoglobin C disease, hemoglobin HbSC disease or, sickle cell disease SC	HbSC or SCD-SC	Hemoglobin S gene is inherited from one parent and hemoglobin C gene is inherited from the other parent. This is usually a milder form of the disease.
Sickle beta thalassemia disease: two major types		Hemoglobin S gene is inherited from one parent and the beta thalassemia gene is inherited from the other parent.
1. Sickle beta zero thalassemia	HbS β^0 -thal or SCD-S β^0 -thal	People with HbS β^0 -thal usually have a more severe form; people with HbS β^+ -thal usually have a milder form.
2. Sickle beta plus thalassemia	HbS β^+ -thal or SCD- β^+ -thal	
Rarer type examples:		In these rarer types a person inherits one sickle cell gene from one parent and one abnormal hemoglobin gene from the other parent.
Hemoglobin SD disease or sickle cell disease SD	HbSD or SCD-SD	
Hemoglobin SE disease or sickle cell disease SE	HbSE or SCD-SE	The severity of these rarer types of the disease varies. For example, while HbSE parallels that of HbS β^+ -thal, HbSO ^{Arab} and HbSC ^{Harlem} often have severe disease symptoms and complications similar to people with HbSS. HbS Lepore and HbSD have milder symptoms.
Hemoglobin SO Arab disease or Sickle cell disease SO Arab	HbSO ^{Arab} or SCD-SO ^{Arab}	
Hemoglobin SC Harlem or Sickle cell disease SC Harlem	HbSC ^{Harlem} or SCD-SC ^{Harlem}	
Hemoglobin sickle Lepore or Sickle cell disease Lepore	HbS Lepore or SCD-S Lepore	

Note: Adapted from: Pass et al. (2000).

Table 2

Sample Questionnaire Items

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- Are you able to talk about having sickle cell disease/trait with a partner?
 - Are you able to talk with your partner about him/her having sickle cell disease or trait?
 - How likely is it that you will accept the risk of having a child with sickle cell disease or trait?
 - How likely is it that you will choose to not have children to avoid having a child with sickle cell disease or trait?
 - How likely is it that you will choose to having testing in pregnancy (like amniocentesis) to determine if the fetus has sickle cell disease or trait?
 - How likely is it that you will choose to have test tube eggs or sperm that are free of sickle cell placed in your uterus (womb) to avoid sickle cell disease or trait?
 - How likely is it that you will choose to use birth control to avoid sickle cell disease or trait?
 - How likely is it that you will choose to have sterilization surgery (tubes tied, vasectomy) to avoid sickle cell disease or trait?
 - How likely is that you will choose to adopt a child to avoid having a child with sickle cell disease or trait?
 - How likely is it that you (your partner/wife) will have an abortion to avoid having a child with sickle cell disease or trait?
 - How likely is it that you (your partner/wife) will end or terminate a pregnancy because of your (your partner/wife) health?
 - Would you accept the risk of your sickle cell disease getting worse if you are pregnant?
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