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Toward Understanding Family-Related Characteristics of Young Adults with Sickle Cell Disease or Sickle Cell Trait in the USA

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Abstract

Aims and objectives—To describe the family-related characteristics of young adults with sickle cell disease or sickle cell trait prior to taking part in a randomized controlled trial on sickle cell reproductive health education.

Background—There is a critical need for educational programs that target the reproductive needs of young adults with sickle cell disease or trait. However, little is known about the family-related characteristics (i.e., demographic attributes and reproductive health behaviors) in which these young adults live.

Design—A descriptive cross-sectional analysis.

Method—At study enrollment, 234 young adults (mean age = 25.9 years, 65% female) completed the SCKnowIQ questionnaire. Descriptive statistics depict the demographic attributes and reproductive health behaviors of young adults with sickle cell disease ($n = 138$) or trait ($n = 96$). For group comparisons, independent t tests or Fisher's tests were used, as appropriate.

Results—Young adults with sickle cell trait had significantly higher education, income, and health insurance than those with sickle cell disease. Both groups believed that sickle cell disease

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Conflict of Interest

The authors declare no conflict of interest.

was a severe condition. A majority of young adults with sickle cell disease (65%) had no children compared to 42% of those with sickle cell trait. Most young adults (85% sickle cell disease, 82% sickle cell trait) were not planning a pregnancy in the next six months and many used condoms, withdrawal, or oral contraceptives.

Conclusions—Socioeconomic disparities exist between young adults with sickle cell disease and sickle cell trait. Future research that advances education about how and when to communicate appropriate genetic risk information to partners and children especially for young adults with sickle cell trait would be beneficial.

Relevance to clinical practice—Awareness of the similarities and differences in the family-related characteristics among young adults with sickle cell disease or trait can allow for more tailored reproductive education.

Keywords

family characteristics; family planning education; family research; genetic counseling; reproductive health behaviors; sickle cell disorders

INTRODUCTION

Reproductive health is an important global concern. The United Nations acknowledges that reproductive health is a multidimensional phenomenon and its components are integrated throughout each of the 8 Millennium Development Goals (Bernstein *et al.* 2006, United Nations 2014). Global reports underscore the urgent need to improve access, education, and develop unique programs about reproductive health that target “special populations” (Bernstein *et al.* 2006, p. 17), and in particular, address the needs of people with hemoglobinopathies, which are largely represented by people with sickle cell disease (SCD) (Modell & Darlison 2008).

The reproductive health of people with SCD or sickle cell trait (SCT) is a major health concern. The World Health Organization (2011) estimates that over 300,000 births of babies with severe hemoglobinopathies occur globally each year and millions of people are affected by SCD throughout the world (Creary *et al.* 2007, Modell & Darlison 2008). In the United States (U.S.), about 100,000 people have SCD and the majority are of African ancestry (U.S. Department of Health & Human Services & National Institutes of Health 2012). An additional 3 million or about 1 in 12 African Americans carry the SCT (U.S. Department of Health & Human Services & National Institutes of Health 2012). Although all people now born in the U.S. have newborn screening for SCD, and infants with SCD generally show signs after the first few months of life, only 16% of people with SCT know their SCT status (Treadwell *et al.* 2006). Many parents are not informed of their children's SCT status following newborn screening, and even if informed, some may not understand the consequences of their children's SCT status in regards to the implications for future reproductive decisions (Taylor *et al.* 2014).

Only a few researchers have studied the family-related characteristics of people with SCD or SCT. Investigators have studied the effects of SCD on family functioning (Thompson *et al.* 1993); parents' communication of SCD or SCT to children (Acharya *et al.* 2009); or

education and counseling related to the genetic aspects of SCD or SCT (Kladny *et al.* 2005, Long *et al.* 2011). Yet information about family-related characteristics of young adults with SCD or SCT is extremely limited. To address this gap, this analysis describes the family-related characteristics of young adults with SCD or SCT at the time of their enrollment in a reproductive health study. We define family-related characteristics as the demographic attributes (e.g., age, sex, education, health insurance status), reproductive health behaviors (e.g., pregnancy plans, birth control use) of an intimate couple, and the participants' current children (e.g., number, genotype status). Description of the family-related characteristics is essential information for nurses and other clinicians who provide prenatal, genetic, and reproductive counseling and education to young adults with sickle cell disease or trait and for researchers who design and evaluate studies focused on genetic counseling and education about reproductive health options.

BACKGROUND

SCD is an inherited autosomal recessive condition that has several different major genotypes (Gallo *et al.* 2010). People with the most common and severe form of SCD (i.e., HbSS) inherit two copies of the sickle cell gene; however, other forms include inheritance of one sickle cell gene and one gene from another abnormal type of hemoglobin (e.g., HbC; Hb beta thalassemia) (Creary *et al.* 2007). As a result, people with SCD have variations in clinical severity. The clinical hallmark of SCD is recurrent episodes of pain and acute illness, and progressive damage to most body organs (e.g., brain, kidneys, lungs, and cardiovascular system) (Creary *et al.* 2007). People with SCT inherit one copy of the sickle cell gene and one copy of the normal hemoglobin gene and are typically healthy and rarely exhibit health problems (John 2010).

There are few available studies on reproductive health behaviors of people with SCD or SCT. Three decades ago, Samuels-Reid and colleagues (1984) examined reproductive patterns and contraceptive use among women in the eastern U.S. with SCD. Questionnaires were completed by 48 women with SCD (mean age = 25.3 years, range = 14 to 46 years) and 80 female controls (mean age = 23.7 years, range = 16 to 40 years) of which 18% were women who had SCT. The mean age for onset of sexual intercourse was 17.7 years for the women with SCD and 17.0 years for the control group. Sixteen percent of the women with SCD were married and they had an average of 1.6 living children (range = 1 – 5 children). Thirty-eight percent of the women with SCD were sexually active compared to 81% of the control group females. Regarding the age of onset for sexual intercourse, similar findings were reported (median age of first sex = 17 years) for adolescents in the U.S. (Finer & Philbin 2013) and worldwide (15 – 19 years) (Wellings *et al.* 2006).

Almost a decade passed following the Samuels-Reid and colleagues (1984) study until Howard, Lillis, and Tuck (1993) interviewed 156 women in north London who had SCD about their contraceptive use. The women were 17 to 53 years of age (mean = 28.4 years). About 65% of women had used oral contraception (45% combination pill, 20% progestin only pill), 19% had used an intrauterine device (IUD), and 17% used injectable progestin (i.e., depot medroxyprogesterone acetate). These results are similar to Samuels-Reid and colleagues' (1984) findings that among women with SCD the preferred method of

contraceptive was birth control pills (39%) and about 15.4% used IUDs. Howard et al. found that 64.2% of women experienced an unplanned pregnancy, which is higher than the 38% unplanned pregnancy rate among women with SCD reported by Samuels-Reid et al. (1984). These findings for women with SCD are in contrast to the estimates that about half of the pregnancies in the United States are unplanned (Finer & Henshaw 2006). Noteworthy, neither Samuels-Reid et al. nor Howard et al. included male participants.

The limited available data leave clinicians and researchers with little understanding about how to address the specific reproductive needs of young people with SCD and SCT despite the multiple calls for counseling and education for this population (Creary *et al.* 2007, John 2010, Yusuf *et al.* 2011). The aim of this analysis was to compare the demographic attributes about the perception of severity of SCD and SCT, history of family relatives with SCD, and reproductive health behaviors (i.e., relationship status, children, pregnancy plans, use of birth control methods) for differences by SCD or SCT status among a sample prior to receiving a reproductive health education intervention.

METHODS

Design

A descriptive cross-sectional analysis was conducted using first visit baseline data from young adults with SCD or SCT who participated in a randomized controlled trial that focused on SCD and reproductive health options. We already have reported qualitative findings about the young adults' perceptions of parenthood and study participation (Hershberger *et al.* 2015), outcomes of pilot studies (Gallo *et al.* 2010, Gallo *et al.* 2013) and the randomized intervention study (Gallo *et al.* 2015, Wilkie *et al.* 2013). The study and procedures were approved by the institutional review boards of the University of Illinois at Chicago and the Ann and Robert H. Lurie Children's Hospital of Chicago.

Sample

The sample included 18 to 35 year old English speaking young adults who reported having SCD or SCT and wanting children in the future. Young adults who were legally blind or unable to have children or complete the study were excluded. To decrease the possibility of data contamination, those who knew or had relatives or friends enrolled in the study were also excluded.

Data Collection

Participant recruitment occurred in multiple venues: adult and pediatric sickle cell clinics at the University of Illinois Hospital and Health Sciences System in Chicago, pediatric sickle cell clinic at the Ann and Robert H. Lurie Children's Hospital of Chicago, local community groups, college student centers, drug and grocery stores, and online social networks. Data were collected at the locations of the participant's choice, including clinical settings, participants' homes, or other public locations such as libraries.

Staff at the sickle cell clinics referred patients with SCD or parents of children with SCD to well-trained and experienced research specialists and participants also personally referred

themselves to the study. After research specialists confirmed the person's eligibility or coordinated laboratory screening to confirm sickle cell condition (SCD, SCT), the research specialists obtained signed informed consents. All baseline data were collected using a pentablot computer. Participants received \$25 in cash for their time and travel expenses at the data collection session.

Data Analysis

We generated descriptive statistics of demographic attributes and reproductive health behaviors for participants with SCD and SCT separately. For group comparisons, we used independent t tests for continuous variables and Fisher's tests for categorical variables. Statistical significance was set at a two-sided alpha of 0.05.

Measure

For this analysis, demographics, perception of severity of SCD and SCT, birth control options and couple partnering items were selected from the SCKnowIQ questionnaire. See Table 1 for example items. We previously published on the development, validity, and reliabilities of the SCKnowIQ (Gallo *et al.* 2010, Gallo *et al.* 2013, Wilkie *et al.* 2013).

RESULTS

Demographics

Baseline descriptive questionnaire items were completed by 234 young adults (65% female, 94% African American or Black) who had either SCD ($n = 138$) or SCT ($n = 96$). The young adults with SCD had a group mean age of 25.7 years ($SD = 4.7$) and the young adults with SCT had a group mean age was 26.1 years ($SD = 5.2$); the difference was not statistically significant ($p = 0.51$). Other sample demographic characteristics appear in Table 2. By sickle cell status groups (i.e., SCD or SCT) significant differences were found for sex ($p < 0.001$), highest education level ($p = 0.004$), family income ($p = 0.005$), employment status ($p < 0.001$), and health insurance ($p < 0.001$). Compared to the SCD group, those with SCT were more likely to be female, to have higher education and income levels, and to have full-time work and employer or union health insurance.

Severity of SCD or SCT and History of Relatives with SCD

When young adults with SCD or SCT were asked to rate the severity of SCD and SCT on a ten-point scale, both groups had similar average scores for either severity of SCD or severity of SCT, but both groups indicated SCD was nearly twice as severe as SCT. For severity of SCD, the average severity score was 9.2 ($SD = 1.5$) for those with SCD ($n = 137$) and 9.0 ($SD = 1.7$) for those with SCT ($n = 94$). For severity of SCT, those with SCD ($n = 135$) indicated an average severity score of 5.4 ($SD = 3.1$) and those with SCT ($n = 93$) indicated an average severity score of 5.7 ($SD = 2.7$). More than half of the sample with SCD ($n = 74$; 54%) had a family relative with SCD and slightly less ($n = 39$, 42%) of the sample with SCT had a family relative with SCD.

Partner and Relationship Status

By sickle cell status groups, a significant difference was found for partner sickle cell status ($p < 0.001$). Many participants indicated that either their partners had normal hemoglobin (SCD: $n = 79$, 58%; SCT: $n = 35$, 38%) or they had no partner (SCD: $n = 38$, 28%; SCT: $n = 20$, 22%). Four (3%) young adults with SCD had partners with SCD and three (2%) had partners with SCT. Only three (3%) participants with SCT had a partner with SCD, however, 18 (19%) had a partner with SCT.

Regarding marital status, a majority of young adults were never married (SCD: $n = 120$, 87%; SCT: $n = 73$, 76%). Ten percent ($n = 14$) with SCD and 18% ($n = 17$) with SCT indicated they were married. A low number of people across both groups indicated they were either separated (SCD: 1%, SCT: 1%) or divorced (SCD: 2%, SCT: 5%).

Status of Children

Among young adults with SCD or SCT, a significant difference was found for total number of children ($p < 0.001$). Whereas 42% ($n = 40$) of young adults with SCT had no children, 65% ($n = 90$) of young adults with SCD did not have children. For those with SCD, 17% had 1 child and 13% had 2 children; only 5% had 3 to 6 children. For those with SCT, 17% had one child, 18% had 2 children and 24% had 3 to 6 children. Whereas young adults with SCD had a total of 83 biological children, those with SCT had 136 biological children. The mean number of biological children was 0.6 (SD = 1.0) for young adults with SCD and significantly lower than the mean number 1.4 (SD = 1.6) for those with SCT ($p < 0.001$).

Among the young adult parents with SCD, 7 children had SCD and 74 children had SCT. Among those with SCT, 44 children had SCD, 43 had SCT, and 48 had normal hemoglobin. No young adults reported adopting a child. One participant in each of the 2 sickle cell groups reported fostering children; a male with SCD was fostering 2 children and one female with SCT was fostering one child.

Participants reported use of reproductive technologies (e.g., donor eggs or sperm) to offset transmission of sickle cell to their future children. Participants were asked, "In the last 6 months, did you (or your partner) have tested fertilized eggs placed in your (or her) uterus so you won't have a baby with sickle cell disease?" Of the 7 young adults (5 with SCD and 2 with SCT) that answered "yes", only 1 participant with SCD indicated that donor sperm was used. Further details about the number of children of young adults with SCD and SCT can be found in Table 3.

Pregnancy Plans and Birth Control Use

Although fairly equal percentages of young adults with SCD or SCT planned to become pregnant in the next 6 months (SCD: $n = 19$, 14%; SCT: $n = 15$, 16%), most participants were not planning to become pregnant in the next 6 months (SCD: $n = 117$, 85%, SCT: $n = 79$, 82%) and a small percentage indicated they were undecided about whether they were planning to become pregnant in the next 6 months (SCD: $n = 2$, 1%, SCT: $n = 2$, 2%). Seven young adults (SCD: $n = 2$, SCT: $n = 5$) reported themselves or their respective partner had their "tubes tied." Of these seven, 5 women had undergone tubal ligation and 2 men's female

partners had undergone tubal ligation. No male participants or male partners of female participants reported undergoing a vasectomy.

No significant differences on birth control methods were found by sickle cell status group (Table 4). Condoms were the predominant birth control method used by young adults in both groups (SCD: $n = 53$, 39%; SCT: $n = 45$, 48%). Many young adults indicated that they do not use any type of birth control method (SCD: $n = 53$, 39%; SCT: $n = 26$, 28%), but about half in each group were not sexually active [of 53 with SCD, 25 (47%) were not sexual active; of 26 with SCT, 15 (58%) were not sexually active]. Other widely used birth control methods among both groups were withdrawal, oral or transdermal contraceptives (birth control pills or the patch), injectable contraceptives (i.e., depot medroxyprogesterone acetate), or IUDs.

Regarding frequency of birth control use, about a quarter of the participants in each group indicated they were using a birth control method consistently (i.e., “every time”) (SCD: $n = 36$, 26%; SCT: $n = 21$, 22%). Of the rest of the sample, some reported they “never” used birth control in the past 6 months (SCD: $n = 26$, 19%; SCT: $n = 17$, 18%), which is slightly more than the number of SCD or SCT participants that planned to become pregnant in the next 6 months (SCD: $n = 19$, 14%; SCT: $n = 15$, 16%). The lowest percentage of young adults reported they used birth control “half of the time” (SCD: $n = 6$, 4%; SCT: $n = 1$, 1%), which is slightly more than the overall percentage of young adults that indicated they were undecided about planning a pregnancy in the next 6 months (SCD: $n = 2$, 1%, SCT: $n = 2$, 2%). Table 5 depicts the frequency of birth control use.

DISCUSSION

Previous research rarely has looked at family-related characteristics of young adults with SCD or SCT of childbearing age. Our analysis of data collected from participants who answered questionnaire items on computers provides another way to understand the family context in which young adults with SCD or SCT live. For example, we found significant demographic differences between young adults with SCD or SCT. Because young adults with SCT surpassed young adults with SCD in high levels of education and family income and also employment and obtaining health insurance, young adults with SCT appear to have more opportunities for higher advancement compared to young adults with SCD who suffer the effects of the disease. There may be several reasons for these demographic differences in this sample. Foremost, other researchers have demonstrated the challenges that adults with SCD have in finding and keeping employment, continuing their education, and advancing their careers due to the physical constraints and instability that accompany a diagnosis of SCD (Thomas & Lipps 2011). Another underlying reason may be that we recruited more women in the SCT group and in recent years young women as a group have outnumbered their male counterparts as college graduates in the U.S. (Buchmann *et al.* 2008). It is unclear why young adults with SCT in our sample had higher educational and family income levels than young adults with SCD, however, the disparity we identified warrants further investigation about the challenges that face young adults with SCD.

What is clear is that young adults with SCD or SCT indicated that SCD was severe and that SCT was moderately severe. These beliefs about the severity of either SCD or SCT likely reflect the community knowledge since about half of the sample had personal experiences with the conditions among their relatives. In our prior qualitative research, we found that people with SCD thought their personal experience with SCD had influenced their decisions to remain childless to avoid passing on the suffering they had experienced to their future children (Gallo *et al.* 2010).

Our findings about young adults' partners and relationship status indicated that most young adults with SCD (87%) or SCT (76%) were never married even though the group mean age was about 26 years. This finding is consistent with a growing trend to delay marriage in the U.S. where current estimates indicate the median age at first marriage is about 26 years for young women and about 28 years for young men (Copen *et al.* 2012). Another reason for the low marriage rate, as voiced by adults at-genetic risk for Huntington's disease, could be that some participants are reluctant to engage in long-term relationships so as not to burden a partner with the consequences that an illness or increased genetic risk can bring (Klitzman *et al.* 2007). Moreover, African Americans tend to marry far less and much later in life (if at all) because of a lack of wealth accumulations (i.e., owning a car or house, having a bank account) and resources (i.e., education) even though they may want to marry (Schneider 2011).

For those young adults that reported having partners, significant differences between the SCD and SCT groups were found when comparing partner's sickle cell status. Most young adults with SCD had partners with normal hemoglobin compared to those with SCT. Whether young adults with SCD purposely sought-out partners that were free of SCD or SCT is unknown. The little research completed about partner selection among people with genetic disease or risk has shown that young adults face a series of dilemmas in dating situations including when and how to disclose their genetic risk or illness to avoid experiences of rejection or stigma (Asgharian *et al.* 2003, Klitzman & Sweeney 2011). Because clinical education has traditionally targeted those with SCD or parents of children with SCD, it may be that these individuals are better educated about transmission of SCD and purposely select reproductive partners that present less risk of transmitting SCD to their future children.

Participants in both the SCD ($n = 48$) and SCT ($n = 56$) groups had biological children, only two participants were foster parents, and none had adopted children. This finding is consistent with other research indicating the importance of biological children in this population (Asgharian *et al.* 2003, Gallo *et al.* 2010). Of the 104 participants that reported having children, 168 children had either SCD ($n = 51$) or SCT ($n = 117$). Among the children with SCD, the vast majority had a parent with SCT ($n = 44$). The accumulating evidence about the desire for biological children points not only to a need for reproductive education to facilitate informed decision making for adults themselves but also an emergent need, especially for young adults with SCT, for providing education about when and how to deliver developmentally appropriate genetic information to prepare their children for future reproductive decisions (Rowland & Metcalfe 2013).

Most participants reported they were not planning to become pregnant in the next 6 months. Among those who did opt to use birth control, condoms were the preferred birth control method used. The high use of condoms (42% of the total sample) could be because condoms provide some protection against sexually transmitted diseases as well as some protection against pregnancy and are typically accessible. The use of withdrawal as a birth control method by about equal numbers of those with SCD (17%) or SCT (18%) is an important finding. The high use of condoms and withdrawal, albeit the low level of protection afforded by these methods against pregnancy (i.e., 17% [condoms] and 18% [withdrawal] probability for failure after 12 months of use, Kost *et al.* 2008), is concerning. It maybe that young adults, especially women with SCD, are hesitant to use other birth control methods that contain hormones that could potentially compromise their health. Yet, earlier reports have shown that 39% to 65% of women with SCD opted to use birth control pills (Howard *et al.* 1993, Samuels-Reid *et al.* 1984), which do contain hormones. In these reports, investigators were specifically focusing on contraceptive use in samples of women that were either 14 to 46 years of age (Samuels-Reid *et al.* 1984) or 17 to 53 years of age (Howard *et al.* 1993), while we were focusing on understanding young adults family-related characteristics at time of enrollment in a reproductive educational intervention, which may account for these differences. Nonetheless, the use of IUDs and depot medroxyprogesterone acetate for birth control in our sample and in the sample reported by Howard and colleagues (1993) are similar. We also found that some participants (39% SCD, 28% SCT) reported they did not use any type of birth control method, however, about half of the young adults in this group were not sexually active.

Despite these insights into family-related characteristics about young adults with SCD or SCT, there are a few limitations about the study and lessons we learned that warrant discussion. We found that seven participants reported they or their partners had undergone sterilization. These young adults should not have been enrolled in the study, but they were honest when responding to the questionnaire items once enrolled. These young adults may have been interested in more education, the compensation, or did not understand the eligibility questions; we are not able to know participants' reasons. Likewise, seven participants indicated they had "tested" fertilized eggs placed in their or their partner's uterus. Yet, only one participant reported the use of donor sperm or other in vitro fertilization procedure. This discrepancy may indicate that some participants at enrollment may not have fully understood all the available reproductive options or fully comprehended specific details about assisted reproductive procedures, which were goals of the CHOICES intervention. We also caution the interpretation of our findings regarding the greater number of young adults with SCT that had children with SCD and had partners with SCT. This finding may be related to the substantial number of SCT participants that were recruited from sickle cell pediatric clinics.

Additionally, our sample was comprised of more women than men. This may be because we recruited at sickle cell pediatric clinics where mothers typically bring their children with SCD for care. However, our related studies on pain have found that more women with SCD opt to participate (Ezenwa *et al.* 2015, Wilkie *et al.* 2010). Much of the sample was obtained from a large, urban area in the Midwestern U.S. Noteworthy is that we do or have resided in this area and acknowledge our personal and professional views that reproductive decisions

among young adults with SCD or SCT should ultimately reside with informed individuals. Other geographic locations where participants have differing cultural views and norms about family and reproduction may have more nuanced or alternative findings. We also acknowledge the increased possibility of a Type 1 error in our results because of the large number of statistical tests carried out. What we have reported in this exploratory study are preliminary findings that could inform future hypothesis driven studies.

These findings point to several important implications for future research. Subsequent research should include more detailed questions about family-related characteristics and the composition of individuals that comprise the family. For example, we are curious about young adults' ages at the birth of their first children, the children's sickle cell status, and whether age at initial parenthood or the child's sickle cell status affected future reproductive decisions. Research that examines whether, when, and how young adults with SCD or SCT choose to communicate their genetic status to future sexual partners and how they find out about their partners' sickle cell status would be beneficial. Although we are one of the first to include men with SCD or SCT in our sample, more knowledge about how men who are at-risk for transmitting SCD to their future children make reproductive decisions would also be beneficial. The need for targeted education not only for those with SCD but for young adults with SCT became evident. Because many of the participants with SCT selected reproductive partners with SCT and these couples had children, the need for designing and implementing developmentally appropriate parent-child educational tools also came to light. The findings have sparked our interest in how and for what reasons young adults choose to use and adhere to birth control methods in the face of SCD or SCT. In view of our results, we are curious about how beliefs about the severity of SCD or SCT effect these reproductive behaviors. Finally, more consideration about how eligibility screening questions are asked and how potential participant responses are evaluated when designing future studies is justified.

CONCLUSION

This study is among the first to report family-related characteristics of young adults with SCD or SCT collected through computer technology. Although most of the 234 young adults in our sample were never married, those with SCT were more likely to have higher levels of education, higher family income, employment, and health insurance than young adults with SCD, which indicates a potentially vulnerable socioeconomic situation for those with SCD. For young adults that reported having a partner, a greater number of young adults with SCD had partners with normal hemoglobin compared to partners of young adults with SCT. However, those with SCT had a higher percentage of partners with SCT compared to partners of young adults with SCD. Partner selection is important among young adults with SCD or SCT as the partners' sickle cell status affects the risk of transmitting SCD to future children and guides clinical education about reproductive options.

More participants with SCT had children and their children were more likely to have SCD compared to the young adults with SCD. Only a couple of young adults had foster children and no participants opted for adoption. These findings provide support for the need for reproductive education that focuses not only on young adults who are likely to want their

own biological children but also on educational efforts that cut across generations especially parent-child interventions. In this sample, only a small percentage were planning to become pregnant in the next 6 months (SCD: 14%, SCT: 16%) and only a few young adults in both the SCD and SCT groups were undecided about pregnancy. The birth control method of choice was condoms (SCD: 39%, SCT: 48%) or withdrawal (SCD: 17%, SCT: 18%) in a sample that included abstinent young adults.

These findings have numerous practice implications especially for nurses and other clinicians who are in key positions to provide education and counseling about transmission of SCD and available reproductive options. Several areas for future research were identified including providing more detailed information about the family-related characteristic of young adults with SCD or SCT such as the mean age at initial parenthood. More knowledge about men's reproductive decision processes, the information needs of young adults with SCT, and how best to assist parents in the delivery of developmentally appropriate genetic information to their children, would also be beneficial.

RELEVANCE TO CLINICAL PRACTICE

Young adults with SCD or SCT recognize that SCD is a severe condition. This finding has multiple implications for nurses and other clinicians who provide education and counseling for these young adults. Foremost, opportunities to proactively engage young adults in discussions about the differences between SCD and SCT and how partner sickle cell status impacts risk of transmitting SCD to their future children would provide a more comprehensive approach to the care of young adults with SCD or SCT. Helping young adults understand the complexities about sickle cell inheritance, the potential health effects, and the implications for their own future regarding informed reproductive decisions are essential. Nurses that provide this information can empower young adults with SCD or SCT with the ability to share accurate information with partners, current or future children, and extended family members.

In relation to family planning, inquiring about plans for future pregnancy and discussing available birth control options are important components of this education. Moreover, nurses in a variety of settings including those practicing in women's health, men's health, and in pediatric clinics are in key positions to address the reproductive needs of young adults especially those with SCT who may not attend sickle cell clinics. Nurses should advocate for policies that would reduce the potential for socioeconomic disparities among both sickle cell groups but especially among young adults with SCD.

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SUMMARY BOX**'What does this paper contribute to the wider global clinical community?'**

- Awareness of the socioeconomic differences and family-related nuances between young adults with sickle cell disease and sickle cell trait can lead to better reproductive education and counseling programs for this population.
- Nurses and other clinicians can provide high quality clinical care by assessing desire for parenthood and, when appropriate, addressing anticipatory guidance about how a partner's or a potential partner's sickle cell status can influence the transmission of sickle cell disease to future children.
- Future research that investigates the family-related characteristics including whether, when, and how young adults with sickle cell disease or trait choose to communicate their genetic status to future sexual partners would be beneficial.

Table 1

Examples of SCKnowIQ Questionnaire Items

- How serious do you think sickle cell DISEASE is on a scale of 0 to 10, with 0 being not serious and 10 being really serious?
- How serious do you think sickle cell TRAIT is on a scale of 0–10, with 0 being not serious and 10 being really serious?
- In the next six months, do you plan to get pregnant by your partner(s)? (woman)? In the next six months, do you plan to get your partner(s) pregnant (man)? [Response options: Definitely no; Probably no; Probably yes; Definitely yes; I don't have a partner]
- In the past six months, HOW OFTEN did you use a birth control method when you had sex with your partner? [Response options: Never; A few times; Half of the time; Most of the time; Every time; I did not have sex; I don't have a partner]
- Do you have a relative with sickle cell disease?[Response options: Yes; No]
- Does your partner have sickle cell DISEASE/TRAIT? (Partner means significant other, spouse, boyfriend, girlfriend, or sexual partner)[Response options: Yes; No; Don't know; No partner]

Table 2Demographic Characteristics by Sickle Cell Group (*N* = 234)*

Variable		SCD (<i>n</i> = 138)	SCT (<i>n</i> = 96)	<i>P</i> value
Sex	Women	74 (54%)	78 (81%)	< 0.001
	Men	64 (46%)	18 (19%)	
Age	Mean (SD)	25.7 (4.7)	26.1 (5.2)	0.514
	18–25	71 (51%)	50 (52%)	1
	26+	67 (49%)	46 (48%)	
Race	African American	62 (45%)	48 (50%)	0.773
	Black	68 (49%)	42 (44%)	
	Mixed Race	4 (3%)	4 (4%)	
	Other	4 (3%)	2 (2%)	
Ethnicity	Hispanic	6 (4%)	4 (4%)	1
	Non-Hispanic	132 (96%)	92 (96%)	
Highest Education Level	High school (9–12)	46 (33%)	32 (33%)	0.004
	Some college or vocational school	77 (56%)	37 (39%)	
	4-year college degree	11 (8%)	17 (18%)	
	Graduate degree	4 (3%)	10 (10%)	
Family Income level	< \$10,000	59(43%)	30(31%)	0.005
	\$10,000 to 29,999	43 (31%)	21 (22%)	
	\$30,000 to \$49,999	14 (10%)	25 (26%)	
	\$50,000 or more	18 (13%)	19 (20%)	
	Unknown	4 (3%)	1 (1%)	
Employment status	Full-time	16 (12%)	31 (32%)	< 0.001
	Part-time	21 (15%)	22 (23%)	
	No, full-time student	18 (13%)	17 (18%)	
	Not employed	83(60%)	26(27%)	
Health insurance	Employer or union	14 (10%)	32 (33%)	< 0.001
	Medicaid/Medicare	103(75%)	45(47%)	
	Private insurance	3 (2%)	1 (1%)	
	No insurance	14 (10%)	15 (16%)	
	Unknown	4 (3%)	3 (3%)	

*Independent t tests were used for comparison of continuous variables and Fisher's tests were used for comparison of categorical variables.

Table 3

Family Characteristics: Children *

	Value	SCD (n = 138)	SCT (n = 96)	P value
Number of children	0	90 (65%)	40 (42%)	< 0.001
	1	23 (17%)	16 (17%)	
	2	18 (13%)	17 (18%)	
	3	6 (4%)	12 (13%)	
	4	0 (0%)	6 (6%)	
	5	0 (0%)	4 (4%)	
	6	1 (1%)	1 (1%)	
Number of biologic children with sickle cell disease	0	132 (96%)	62 (65%)	< 0.001
	1	5 (4%)	25 (26%)	
	2	1 (1%)	8 (8%)	
	3	0 (0%)	1 (1%)	
Number of biologic children with sickle cell trait	0	93 (67%)	67 (70%)	0.245
	1	22 (16%)	20 (21%)	
	2	18 (13%)	6 (6%)	
	3	4 (3%)	2 (2%)	
	4	1 (1%)	0 (0%)	
	5	0 (0%)	1 (1%)	
Number of biologic children with normal hemoglobin	0	138 (100%)	65 (68%)	< 0.001
	1	0 (0%)	18 (19%)	
	2	0 (0%)	9 (9%)	
	3	0 (0%)	4 (4%)	
Total number of children (Biological and Foster)		83	136	
Number of children per person (mean ± standard deviation)		0.6 ± 1.0	1.4 ± 1.6	< 0.001
Total number of children with SCD		7	44	
Total number of children with SCT		74	43	
Total number of children without SCD/SCT		0	48	
Total number of children with unknown SC status		2	1	
Adopted children	No	138 (100%)	96 (100%)	1
	Yes	0 (0%)	0 (0%)	
Foster children	No	137 (99%)	95 (99%)	1
	Yes	1 (1%)	1 (1%)	

* Independent t tests were used for comparison of continuous variables and Fisher's tests were used for comparison of categorical variables.

Table 4Birth Control Use by Method^{*}

Birth Control Method ^a	SCD (n = 137)	SCT (n = 94)	Total Percent	P values
Condoms	53 (39%)	45 (48%)	42%	0.178
None	53 (39%)	26 (28%)	34%	0.092
Withdrawal or “pulling out”	23 (17%)	17 (18%)	17%	0.860
Birth control pills or patch	14 (10%)	16 (17%)	13%	0.163
Depot medroxyprogesterone acetate	17 (12%)	12 (13%)	13%	1
Intrauterine device (IUD)	8 (6%)	11 (12%)	8%	0.144
Emergency contraception (“morning after pill”)	6 (4%)	3 (3%)	4%	0.742
Rhythm, cervical mucous, temperature, or ovulation calendar	3 (2%)	3 (3%)	3%	0.689
Female condoms or vaginal pouches	4 (3%)	1 (1%)	2%	0.651
Cervical cap	2 (1%)	2 (2%)	2%	1
Diaphragm	2 (1%)	1 (1%)	1%	1
Jelly, cream, foams, or sponges	0 (0%)	1 (1%)	0%	0.407

^{*} P values were obtained using Fisher’s test.

^aMissing 3 from larger sample (1 SCD, 2 SCT) due to laptop theft during data collection.

Table 5Frequency of Birth Control Use^{*}

Options for Reporting Birth Control Use ^b	SCD (n = 136)	SCT (n = 94)	Total Percent of both SCD and SCT groups	P value
Every time	36 (26%)	21 (22%)	25%	0.221
Most of the time	18 (13%)	19 (20%)	16%	
Half of the time	6 (4%)	1 (1%)	3%	
A few times	12 (9%)	15 (16%)	12%	
Never	26 (19%)	17 (18%)	19%	
Not sexually active	38 (28%)	21 (22%)	26%	

^{*} Fisher's test was used to obtain the p value.

^b Missing 4 from larger sample (2 SCD, 2 SCT) due to laptop theft (1 SCD, 2 SCT) and missing data (1 SCD) during data collection.