Managing Sickle Cell Disease in Low-Income Families

From their inception, genetic screening programs were viewed as a way to prevent the transmission of hereditary diseases through education, screening, and selective reproductive decision making. The health-belief model suggests that health behaviors are shaped by perceptions of susceptibility to and severity of illness, the benefits and barriers involved in seeking medical care, and cues to actions (Becker 1979). Applied to SCD, this model implies that a diagnosis of sickle cell trait or SCD would underscore one's susceptibility to passing the disease on to a child and would lead to an alteration in reproductive behavior if the benefits of doing so outweighed the costs. Many health officials hoped that early detection of SCD would lead Blacks to exercise caution in childbearing behaviors; as evidenced by Johnson, who, condemning the neglect of SCD, wrote: “With the availability of a simple test for sickle cell trait carriers that makes mass screening possible, sickle cell disease could well become the first hereditary illness to be controlled by genetic counsel-
ing" (1984, 64). This optimistic assertion suggests that SCD education and screening programs can ultimately reduce or end the spread of the disease. But while screening programs have substantially lowered the genetic transmission of some diseases, such as Tay Sachs, we have little evidence that this has been the case with SCD. Among the reasons why this is so are that childbearing patterns, procreative attitudes, and ideas about adequate health among many Blacks often reflect cultural norms that differ from those of the dominant society. Also, while SCD screening programs have remedied some of the racially motivated neglect of SCD, these programs have not dealt with the inequality imposed by gender and class. The low-income African American women in this study valued their right to have children but were often unable to persuade their male partners to be tested for the sickle cell trait. In addition, they often lacked access to the health care that would have offered them such options as prenatal screening.

Mothers in this study understood the contradiction between having SCD medical knowledge and having a child with the disease. Asked to describe their previous knowledge or family history of SCD, mothers who knew about SCD or had been diagnosed with the sickle cell trait related how they came to have a child with SCD without being explicitly asked their reasons for doing so. They explained that their SCD knowledge did not prevent them from having a child as a result of medical mismanagement or family secrecy. Both explanations emphasize inadequate SCD information, and they are mutually reinforcing because medical mismanagement appears to promote stigma and misinformation and thus family secrecy about the disease. The medical mismanagement theme, especially in terms of the historic neglect of SCD, was frequently voiced by political activists of the 1970s, and much of this rhetoric became part of the appeal to Blacks to participate in SCD screening programs. Ironically, some SCD programs, hastily and poorly organized, furthered the medical mismanagement of the disease by disseminating misinformation and making erroneous diagnoses.

After having a child diagnosed with SCD, mothers embraced the political stance on SCD advanced by those activists who argued that the disease had become a major health problem because it had been ignored by the medical profession for so long. Mothers saw themselves and their children as victims of the historic neglect of SCD, and their own experiences with SCD screening programs support the claim that such programs often failed to provide adequate medical knowledge about SCD or correct diagnoses. Consistent with this stance, most mothers embraced education as the key to preventing the genetic transmission of SCD. Based on their statements, one would expect mothers to seek or accept accurate SCD information, promote SCD screening among members of their own families, persuade their male partners be tested for the sickle cell trait, and engage in selective reproduction to avoid passing the trait on to the child. Mothers in this study, however, rarely did any of these things; thus their actual behaviors contradicted their advocacy of education as a viable strategy for controlling SCD. Clearly, education about SCD did not lead most of these mothers to avoid the risk of passing the disease on to a child. Instead, structural barriers to health care, gender inequality
between these women and their male partners, and the cultural ideology of motherhood all served to undermine the power of education to effect change.

Virtually no research has been done on how having the sickle cell trait affects the reproductive decisions of Blacks, and this study of one group of low-income women who chose to become mothers despite having the trait cannot be generalized to all Blacks. This study does, however, allow us to begin to understand why some low-income Black women may lack the inclination or power to comply with the subtle mandates of SCD screening programs. Those who have the sickle cell trait or SCD have three reproductive options if they are to avoid passing the disease on to a child: They can have their partners tested for the trait, they can undergo prenatal diagnosis of SCD and selective abortion, and they can forgo parenthood. These options assume that pregnancies are planned and that they occur within the context of marriage, that women have enough power in their relationships with male partners to insist they be screened for the trait, that adequate and early prenatal care is available, that women have pro-choice abortion attitudes, and that motherhood is one life option among several, rather than a central role and identity. The life circumstances and values of the mothers in this study did not coincide with these assumptions. The only feasible option for most mothers who wanted to be sure not to pass the disease on to a child was to forgo motherhood, thus relinquishing one of the few areas of life over which they exercised control and from which they derived satisfaction. A diagnosis of SCD posed a threat to the reproductive autonomy of these mothers; they responded by denying, confounding, or doubting SCD medical knowledge, instead of accepting it.

Medical Mismanagement

The mothers spoke of medical mismanagement of SCD in explaining the contradiction between having medical knowledge about SCD and having a child with the disease, a position well supported by studies of SCD screening programs and mothers’ own experiences. Physicians have lacked proper medical knowledge about SCD, screening has not been performed on a timely basis or efficiently, and results have often been erroneous. Much sickle cell testing has not detected hemoglobin C or thalassemia traits, resulting in the births of children with S–C or S–thalassemia. Those with SCD sometimes have been diagnosed as having only the trait, thus suffering illness symptoms for long periods before being accurately diagnosed. And, as studies consistently have documented, “poor patients tend to be grossly uninformed about the details of what was happening to them, mostly because the health care professional viewed them as uneducated and incapable of understanding what was happening” (DiMatteo and Friedman 1982, 140–141).

The legacy of medical mismanagement experienced by mothers promoted mistrust of medical authority and wisdom. These mothers understood the racial politics surrounding SCD and made numerous allusions to the fact that SCD had not been adequately studied because it was thought to affect only Blacks:

When [my niece] had it, it was real weird, because they hadn’t studied it at all then. They told us that we needed to give her more milk, that milk was one way of treating it—Milk! They didn’t really know. And [the doctor] now says
that since it’s a black disease, they never had really looked into it.

Medical mismanagement led mothers to doubt not only the authority but also the intentions of health-care experts. One mother described a “blood problem” her father had that caused symptoms almost identical to those of SCD but that was never diagnosed as SCD. Another mother of two sons with SCD noted that their symptoms resembled those experienced by two of her siblings as she was growing up but that they were never diagnosed with SCD:

I had a brother who had a lot of sickness, and some of the same reactions that [my son] has had, but they called it polio. I have a sister who has a child who has never walked, and they diagnosed him as having muscular dystrophy. It could be, but when my mother found out about [my son] she began to reminisce about these things, and how some of these children had reacted, and some of the problems she’d had with my sisters and brothers. It made me understand better what SCD was ... but prior to 1970 and 1969, you just didn’t hear a lot.

For some mothers, a belief in medical neglect of SCD was replaced by a fear that physicians were labeling too many Black children as having SCD. These mothers felt that physicians were likely to overemphasize SCD, to diagnose the disease when it was not present. One mother, whose nine-year-old daughter was initially diagnosed as having SCD when she was three, said she ignored the diagnosis and her daughter’s symptoms for nearly two years because she believed the diagnosis was racially motivated. To her, it seemed that “they were just going crazy with sickle cell” during that time:

I was always taking her back and forth to the hospital for checkups and stuff; she just had colds and stuff like that. But when she got to be five years old ... she came running home from school one day crying that something in her head [was hurting]. So when I really accepted it, she was five. They had first said it when she was three, but I said no way!

Evidently, some instances of the denial noted among those diagnosed with SCD may well have stemmed from their experiences with screening programs.

Family Secrecy

Health-care experts and mothers agreed that before the SCD screening efforts in the 1970s, secrecy, stigma, and silence often surrounded the topic of SCD. Members of the previous generation usually suffered a long time before being told that the disease they had was SCD; even then, there were many misconceptions about SCD. Thus those who received the diagnosis were hesitant to discuss it with others, even with family members. One thirty-two-year-old respondent said that her own mother anguish over who in the family had the trait for nearly eight years before finally getting tested. Her mother’s sense of responsibility for passing SCD on to her grandson diminished her ability to be supportive. The parents of mothers who knew about SCD were not likely to discuss the disease with their children. One respondent, who grew up in a family of twelve children, had an older sister who suffered from severe pain throughout childhood and
was diagnosed as having SCD in early adulthood. She did not learn that her sister had SCD until her own son was diagnosed as having it. Her mother then confided that her sister had the same thing:

*I don’t think [my mother] really understood, although she had a daughter with it. She didn’t have to deal with it because it was later in life when she was diagnosed. That’s the way I remember it. I never knew until [my son] was born that she had it. She was my oldest sister, and I don’t remember very much about what she went through. [SCD] hadn’t been around all her life, so she was older when she was diagnosed. She was having the pain and didn’t know what it was.*

Another mother implicated her mother-in-law as responsible for keeping the SCD in the family a secret. This mother knew she had the sickle cell trait but was convinced that her husband, who had spent several years in the military, did not have the trait. When their third child was born with the disease, her husband, insisting he had had many blood tests while in the military, denied paternity. His mother then admitted that she and three of her four children had the sickle cell trait.

Although mothers in this study tended to discover a family history of SCD only after their own children had been diagnosed with the disease, they deny that they feel stigmatized about having a child with SCD or are embarrassed to have the diagnosis made known to others. A twenty-one-year-old mother who knew she had the trait described her response this way:

*I was sick. I was sort of mad at myself because I took that chance. I cried. But that was it. [How do you feel now?] Well,*

*I really still haven’t accepted it—I mean, I’ve accepted it more than I did at first, because at first I was just going around saying that she just had the trait, even though I knew better. Now I go ahead and admit she has it. [Is there anything you would ever try to hide?] No. [So this is nothing you would ever try to hide?] No, except for when I was trying to believe that it wasn’t true. I wasn’t ashamed; it was just something that I didn’t want to face at that time.*

Today, more accurate information on SCD may have decreased some of the stigma attached to the disease. But SCD screening programs do not give Black women viable reproductive options for preventing the transmission of the disease.

**Reproductive Alternatives**

By invoking themes of medical mismanagement and family secrecy, mothers persuasively attributed some of the blame for their children’s disease to sociomedical factors—inadequate SCD knowledge, past misdiagnoses, mistrust of the medical system, and the unwillingness of relatives to talk about the disease. Yet once a definite diagnosis of SCD had been made, mothers were confronted with the reality of having the sickle cell trait and the decision whether to have additional children. Despite past deficiencies in SCD genetic screening programs, the trait and disease now can be accurately diagnosed, and SCD medical knowledge is available. In fact, mothers whose children receive care at Children’s Hospital meet with a geneticist during each routine comprehensive health-care visit. Much of the stigma that once sur-
rounded SCD has vanished, and with greater access to medical services and social support, these mothers are in a good position to handle SCD in an open and constructive fashion. My informants suggest, however, that mothers unwittingly perpetuate misinformation about SCD because medical knowledge about SCD challenged the legitimacy of their childbearing behaviors.

Neither public education and screening programs for SCD nor being diagnosed as carrying the sickle cell trait led mothers to alter their reproductive behaviors. But the reality of having passed SCD on to a child made it impossible for these mothers to ignore the reproductive implications of having the sickle cell trait. To avoid the birth of additional children with SCD, they had to make decisions about genetic screening of potential fathers, prenatal diagnosis and selective abortion, and forgoing motherhood. Although these reproductive options were voluntary, evidence shows that members of the dominant society viewed them as appropriate responses to the risk of having an abnormal child or passing a hereditary illness on to a child (Katz Rothman 1986; Rapp 1988). That proper health care and access to these options was available, however, and that motherhood was one of several potentially fulfilling options available to women, simply was not true for these mothers. Motherhood is often an alternative life strategy for low-income black women (Burton 1990), as well as a central identity and source of importance and power. Thus there was an inherent conflict between avoiding the risk of passing SCD on to a child and the ideology of motherhood.

Studies of Tay Sachs screening programs reveal that women play a crucial role in the success of genetic screening programs: they are twice as likely as men to be screened (Beck et al. 1974) and are much more likely than men to insist that their partners be screened (Schneiderman, Lowden and Rae-Grant 1977). The mothers in our study were also more likely than fathers to know they had the sickle cell trait, as it appears they were more responsive to the call to be tested for the disease and more likely to have been screened without giving their explicit consent. Once diagnosed, they were placed in the position of having to persuade their male partners to be tested. This usually met with little success. A twenty-eight-year-old mother of one son discovered she had the sickle cell trait in early childhood, when screening was done at her preschool. Since she knew the implications of having the trait and had planned her pregnancy, I asked if she had asked her boy friend to be tested:

Yeah, but being young and naive, I asked him if he carried the trait, because they taught you when I learned it that if you carry a trait and you marry and have a baby by someone who carries the trait, there's a possibility that the child could come out with the disease. So I just asked him thinking he was tested as a kid like I was. And he told me, "I don't have no trait!" and I went with that. But then when [my son] was born, that's when he found out.

A twenty-two-year-old mother said:

I kept telling my boy friend to get tested, and he said he didn't have anything like that. [But was he actually tested?] Well, he said there was nothing wrong with him, that no one in his family ever had sickle cell anemia and he knew he was healthy. But he did have a niece with it. [So he was never
tested?] No. [And you decided to have a baby anyway?] Well, he sort of wore me down. He looked real healthy, and he was so sure that he did not have the trait. [What did he say when you found out your son has SCD?] He cried. I think he really felt kind of guilty.

Although Black parents react with guilt and depression after having passed SCD on to a child (Evans, Burlew, and Oler 1988), Black men commonly continue to insist that they do not have the sickle cell trait (Travis 1978). The mothers in this study, including some who were married to the biological father of the child with SCD, had difficulty in convincing the child’s father that he had the trait. In fact, some fathers continued to deny having the trait for years, as indicated by one mother whose daughter with SCD is now fifteen years old. Childbirth outside marriage and the gender imbalance of power in these relationships nullified the efforts of Black mothers to have their male partners tested.

Although prenatal diagnosis of Tay Sachs disease was available to the Jewish community before screening programs began, this technology was not available for SCD until the mid-1970s (Mulvihill, Walters, and Wertz 1989) and still is not a routine part of prenatal care for most women with the sickle cell trait (Whitten 1992). Low-income African American women often receive late and/or inadequate prenatal care in the public sector of the health-care system, and as many as 22 percent receive no prenatal care at all (Reed 1990). Many mothers in this study were screened before SCD could be prenatally diagnosed and, given cultural and structural obstacles to obtaining health care, were not aware that such technology existed. Prenatal diagnosis of SCD is now technically available. P. T. Rowley (1989) summarized prenatal screening outcomes at twelve regional centers between 1978 and 1987. He found that 39 percent of prenatal diagnoses of sickle cell disease and 23 percent of prenatal diagnoses of S–C disease resulted in termination of the pregnancy. Although comparisons are difficult, this suggests that a prenatal diagnosis of SCD is less likely to result in abortion than the prenatal diagnosis of other diseases or defects (Katz Rothman 1986).

Only one mother in this study was offered the option of prenatal screening and selective abortion for SCD. She had a child with SCD who had been seriously ill for more than two years before SCD was diagnosed; by that time, this mother was pregnant with a second child. Her son’s long and “mysterious” illness had placed quite a strain on her marriage, and the doctor offered to screen the unborn child for SCD. This mother, still unsure about the exact probability of having another child with the disease, refused the option, and her second child was also born with SCD:

They told me there was a test they could do while I was pregnant and determine if the child had SCD. I had the choice of aborting the pregnancy, but I decided not to let them do the test. I wasn’t really sure of the diagnosis of one out of four, that it was that great again with a second child, that there was a 25 percent chance that the child will have it. I wanted to go ahead, since I was pregnant, and have it, so I didn’t have the test.

All the mothers in this study said they would not have had an abortion to avoid the birth of a child with SCD, although it must be noted that in reality the abortion question never
arose, since they had never been offered the option. One mother said she would not want to “put another child through having SCD” and brought up prenatal testing as a way to make sure it did not happen. Asked directly if she would have an abortion if she knew her unborn child had SCD, she said: “Oh, no! I’d have the baby. He didn’t ask for [SCD].” Another mother introduced the topic of abortion as a way to avoid having a child with SCD but then decided that she could never have an abortion:

*Because they have a thing now where you can be tested in the early months of pregnancy, and I think it’s like within the first three months. And they can determine whether the child has [SCD] or not . . . but I don’t think I would have had an abortion because I don’t believe in them. So, you know, I just would have been praying that he was one of the lucky ones, or that it wasn’t severe, you know, or something.*

These findings parallel the more extensive research of Rayna Rapp (1988), who also found SCD patients less willing than others with genetic traits and disorders to use amniocentesis or choose abortion. These attitudes contradict research showing that Black women have higher rates of abortion than white women do, but there is some evidence that Black and white women tend to abort under different circumstances. K. Trent and E. Powell-Griner (1991) reported that while Black women have higher rates of abortion than white women, Blacks are less likely to abort than are whites if they are teenagers and unmarried or are in their first pregnancy. Persons with lower education are also less likely to abort than more educated persons are. Thus the attitudes of mothers in this study may reflect the fact that they were young and unmarried at the time of their first pregnancy and had low levels of education.

Forgoing motherhood was an option for the thirteen women who knew they had the sickle cell trait before becoming pregnant, and for all the mothers after one child had been diagnosed with SCD. Yet eighteen of these mothers had additional children; those mothers who had no additional children after the SCD diagnosis were either open to the possibility of having more children or had already reached their desired family size. Only three mothers with adequate SCD information admitted that they understood the genetic implications of SCD but took the risk. One mother of a child with SCD said:

*They did tell me to kind of think about having another one, to be careful. Because I just had too many things going against me, and then with her father and me both having SCD, it was kind of a risky deal. So he [the doctor] told me to kind of think about it before I had any more. And I had one more, and that was it. But she came out with the sickle cell trait.*

The majority of mothers, however, did not admit that they took the risk. Rather, they blurred the medical facts about SCD to justify their reproductive decisions.

**The Obfuscation of SCD Knowledge**

While medical mismanagement and family secrecy are credible explanations for the past reproductive behaviors of mothers, I observed that mothers had many opportunities to acquire and use SCD knowledge, but did not take advantage of them. K. Charmaz (1988) has noted that in qualitative
research one must examine carefully what was said as well as what respondents "lack, gloss over, or ignore." The gaps in mothers' stories—what was not said—shape my argument that mothers obfuscated medical knowledge. Although early SCD programs were often deficient, these mothers now have access to accurate SCD information (a regional clinic on SCD) and opportunities for free and voluntary screening. Yet mothers unwittingly contribute to the lack of SCD knowledge by ignoring, doubting, or redefining the medical facts about the disease.

The responses of most mothers suggest that if they had been given adequate SCD knowledge, they might have been more cautious in their reproductive behavior, but their subsequent reproductive behavior refutes this suggestion: eighteen (56.2 percent) of the thirty-two mothers became pregnant again after having one child diagnosed as having SCD. Those with less SCD knowledge before the diagnosis of the first child were somewhat more likely to have additional children. The social position and childbearing patterns of the mothers left them with only one real option for preventing the genetic transmission of the disease, but forging motherhood was not acceptable for the low-income Black women in this study. They placed a central value on the role of motherhood. Confronted with the medical facts of SCD, mothers used various strategies to minimize or nullify the impact of having the sickle cell trait on their lives.

In some cases, mothers spoke persuasively about the need for SCD education, yet their childbearing decisions seemed unaffected. One mother, diagnosed with the sickle cell trait as a teenager, had three spontaneous abortions (fetal wastage is characteristic among persons with the trait) before, at age twenty-seven, carrying a child to full term. She and the child's father were especially disappointed at the diagnosis of SCD. Despite her early diagnosis as a sickle cell trait carrier, she said never understood the consequences of it:

*I didn’t know that if you came into contact with another person with sickle cell, and you and him had a trait, you had a 25 percent chance of having a baby with it. I didn’t know that until afterward. Nobody told me by having the trait you have to watch who you came into contact with. Nobody never explained that to me. So how was I know that he had the trait and I had the trait, and my baby would have the disease?*

One might assume from her statement that the SCD knowledge she now has will affect her childbearing patterns, but this does not appear to be the case. She has been pregnant since the diagnosis of SCD and has experienced another spontaneous abortion. Still, she hopes that she and her husband will have another child.

One twenty-four-year-old mother was screened for the trait during her teenage years and knew that her mate needed to be screened. She says he erroneously tested negative for the trait, and she thought it was safe to have a child:

*They said if I ever get pregnant I had to have my partner tested to see if he had the trait, or something to that effect. But he was tested, but they didn’t test him for hemoglobin C trait, so we didn’t find out that he had the hemoglobin C trait until after [my son] was diagnosed.*

Despite her implication that adequate SCD knowledge would have resulted in more caution, she has had another child with the same mate.
A twenty-two-year-old college student neutralized SCD medical knowledge by pointing to contradictions in statistics on the probability of two parents with the trait passing the disease on to a child. The implication is that even medical professionals really do not know the chances of having a child with SCD:

What do they say about having other children? [Well, there’s a 25 percent chance with each pregnancy that the child will have SCD.] I hear so many things about that—somebody told me there was a 50 percent chance, someone else said a 30 percent chance. Your statistics are the fourth set I’ve heard, with people drawing out little charts! So I don’t think they know.

The most frequent reason mothers gave for not worrying about having another child with SCD was that subsequent children had a different father. Their statements, however, made it clear that in many cases they simply assumed that it was unlikely for them to become involved with another man with the sickle cell trait. As one mother stated, she knew “for sure” that she would “probably” have not taken the same chance again:

They have two different fathers. So I know for sure that I probably would never have taken that chance again, if I were still with [the same man]. But I didn’t think it would happen again.

One mother said that after the birth of a child with SCD, both she and a second child’s father were tested before she became pregnant again. Yet she continued to deny that she had the trait, which casts doubt on whether she was actually tested or is simply assuming that her current partner does not have the trait:

They didn’t have the same father. And before I had any more children, I made sure that whoever my sexual partner was, you know, her father at that time, we were tested. We got tested. [But you said that you don’t have the trait?] Right. And he didn’t either. Because I knew that I did not want to have another child with sickle cell. I wouldn’t do that to myself, and I wouldn’t do it to the child; it’s not fair.

Of the eighteen mothers who had additional children, four passed SCD on to another child; they expressed few regrets. Their responses reflect the more narrow definitions of good health and the greater tendency to tolerate illness symptoms often found among low-income persons. For example, one mother who knew she and her partner had the sickle cell trait and took the chance, felt she was “lucky” because only two of her four children had SCD:

I thought about it, but not too much. ’Cause they told me that it was chance that other kids might have it, but only one had it. The other kids didn’t have it. I guess I was lucky.

The burden of caring for two children with SCD did lead one mother to alter her reproductive plans. Asked if she would have had additional children if her second son had not had SCD, she said:

Yeah, that was my plan. But I thank the Lord that He didn’t allow that to happen. But I had planned on having five or six kids, six at the most. After [my second son] I began to ask questions about other children. I talked with my own GYN
and some other doctors, and they explained the risk. So I decided not to; I just couldn’t see putting another child through that. So I decided to have a hysterectomy so I wouldn’t have other children. And Lord knows, these two have been all I could handle.

Although fourteen of these mothers had no additional children, few attributed the decision to SCD. The response of one mother, asked if she planned to have any more children, revealed she was more concerned with family life than the issue of SCD:

Well, I don’t know. I have some things I want to do for myself, plus I’ve always wanted all of my children to have the same father, but I’m not with my son’s father anymore. Me and my brothers have the same father, and I’ve always wanted that for my kids, so one man wouldn’t be picking up one child, another man doing something special for another child. But I don’t know.

Overall, these mothers’ attitudes reflect a strong desire to have children and defend their reproductive rights. The importance of motherhood among low-income Black women has been documented in other studies (Anderson 1991; Burton 1990; Collins 1987; Ladner 1971), yet there is virtually no systematic research on attitudes toward parenthood among more affluent Blacks or the impact of having the sickle cell trait in their reproductive decisions. And while there are probably class-based differences in attitudes and options among Blacks with the sickle cell trait, it seems likely that the mothers in this study will pass on to their children their own reproductive attitudes. One mother of two children with SCD

was very much opposed to the idea of cautioning them against marrying someone with the trait. Asked if she would be concerned about her son marrying someone with the sickle cell trait, she said:

No, I wouldn’t! I’m sorry, I wouldn’t. I feel like if that’s what he wants to do, and he knows what he’s been through, and he loves this girl, and wants to get married, and they had twenty of them with it, I mean, that’s up to them. Why should they say, “I love you but I can’t marry you because of sickle cell?” To me, that’s wrong. You know what this person has got, and you know what you have got. You done been through it, and so have them—for all you know, there may be a cure a month after you marry. Or even if a cure never comes up, why marry someone else? Life is what you make it.