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Feasibility of a Nutrition Training Program on Improving Self-Efficacy and Life Skill Development for Adolescents with Sickle Cell Disease

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FEASIBILITY OF A NUTRITION TRAINING PROGRAM ON IMPROVING SELF-EFFICACY AND LIFE SKILL DEVELOPMENT FOR ADOLESCENTS WITH SICKLE CELL DISEASE

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Dissertation Approval

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Abstract

Adolescents with sickle cell disease (SCD) are at risk for growth and nutritional deficiencies due to poor dietary intake. Presently, no efficacious behavioral intervention exists to address the nutritional need in SCD management. The Good Nutrition 4Me Living with SCD program is a multi-component intervention, developed to improve knowledge base and the dietary habits of adolescents living with SCD. This report reviews the outcomes for 5 adolescents that participated in this 5-week program. Dietary intake was monitored daily via text messaging and progress with goals was reviewed weekly in the group sessions. Parents were also updated on their adolescent's progress with dietary goals during a family session in the home setting. Resulted indicated promising effects across participants, with trends towards improvement in knowledge about nutritional recommendations and dietary intake. The Good Nutrition 4Me Living with SCD program was viewed as feasible and palatable, as all evaluable families completed the intervention and one-month follow-up activities.

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Chapter One: Statement of the Problem

Statement of the Problem

The World Health Organization defines health as the state of physical, emotional, and social well being, and not only the absence of disease (World Health Organization, 1948). However, it is estimated that more than 7.5 million children in the United States attempt to maintain healthy development while living with one or more chronic diseases (Jones, 2001). Chronic diseases are defined as diseases that have symptoms with a protracted course and with the involvement of one or more major organ systems (Brown & Marcias, 2001). Some of the most common chronic diseases that occur in childhood include asthma, autism, cerebral palsy, congenital heart disease, cystic fibrosis, diabetes, leukemia, and sickle cell disease (Miceli, Rowland, & Whitman, 1999).

Sickle cell disease refers to a group of hereditary disorders affecting the hemoglobin and the production of red blood cells (Ballas, 2002). The discovery of the sickle hemoglobin by Linus Pauling in 1949 is credited for the establishment of the field of molecular medicine (Bunn, 1997). Later medical advances have broadened the definition of the disease to include a group of disorders including sickle cell anemia (Hb SS), sickle hemoglobin C disease (Hb SC), and sickle beta-thalassaemia (Hb Sβ) (Cummins & Anie, 2003). In the United States, it is estimated that 1 in every 500 African-Americans has sickle cell disease (Chen, Cole, & Kato, 2004; Gil, Carson, Porter, Scipio, & Bediako, 2004). Although advances in medical practice have expanded the average life expectancy for patients with sickle cell disease to 50 years, there is no cure and the treatment of sickle cell disease is palliative at best (Claster & Vichinsky, 2003; McCrae & Lumley, 1998). The prevalence rates of sickle cell disease in the United

States and expanded life expectancy for this population highlights the need for psychologists to become knowledgeable about sickle cell disease, its psychological sequelae, and recommended psychological interventions to support overall disease management (Collins, Kaslow, Doepke, Eckman, & Johnson, 1998).

The predominant symptom of sickle cell disease is acute pain which results from blockage of red blood cells, also known as a vaso-occlusive crisis. Often, the focus of treatment for sickle cell disease is symptom management, but current research is also investigating ways of preventing vaso-occlusive crisis episodes. Some preventative treatments aimed at reducing the probability of vaso-occlusive crisis include transfusion therapy, hydroxyurea, and bone marrow transplantation. Although these interventions have been successful for some individuals living with sickle cell disease (Claster & Vichinsky, 2003), these interventions are invasive and have considerable risks associated with use. As an alternative, current research has been investigating the role of nutrition in the management of sickle cell disease and the ways in which enhanced nutrient intake could prevent or reduce the frequency of vaso-occlusive crisis (Nelson, Zemel, Kawchak, Barden, Frongillo, Coburn, Ohene-Frempong, & Stallings, 2002). Many health related behavioral patterns, including eating habits, are formed in adolescence. Accordingly, interventions focused on altering the dietary habits of people with sickle cell disease would be most advantageous for adolescent patients.

Children and adolescents with sickle cell disease must navigate through the same developmental stages as their healthy counterparts. Adolescence is a critical period for identity formation, characterized by increased efforts for autonomy and individuation from the family of origin. While adolescents living with sickle cell disease are going

through the same struggles as their healthy peers, they must also learn key components of their disease management for optimal transition to an independent adulthood. Important elements of self-care, including adequate dietary intake and regular sleep cycles, are elusive to many adolescents, but these healthy behaviors are essential to the management of sickle cell disease. Consequently, the comprehensive care of adolescents living with sickle cell disease must shift from a passive focus on ameliorating symptoms as they arise (e.g. vaso-occlusive crisis episodes) to a much more active disease management process that includes education on preventative steps and healthy self-care practices that patients can adopt to reduce the likelihood of some symptoms.

Purpose of the Study

The purpose of the present study is to examine the feasibility of a nutritional training program in the management of sickle cell disease among adolescents preparing to transition to adult healthcare settings. It evaluates the role that patient education on nutrition will have on the dietary intake of adolescents living with sickle cell disease. The present study also explores the role of attributional styles, particularly one's perspective on locus of control and self-efficacy, as a potentially mediating factor to compliance with dietary recommendations. It is proposed that participants in the nutritional training program would demonstrate an increased knowledge base about sickle cell disease and the role of nutrition in overall disease management, and also demonstrate improved dietary intake of recommended daily allowances across the five essential food groups. It is also proposed that participants demonstrating an external locus of control would demonstrate an increased knowledge base but no significant change in dietary intake. Lastly, it is proposed that participation in this brief, cognitive-behavioral group

intervention would positively improve disease specific self-efficacy and participants' attitudes towards their illness.

Relevance to Clinical Health Psychology

The discipline of clinical health psychology has expanded in the past 20 years with the growing recognition that psychological issues play a crucial role in most medical conditions. It is estimated that the management of chronic health conditions accounts for approximately 68% of the total health care expenditures in the United States (Llewelyn & Kennedy, 2003). Clinical health psychologists are able to offset some of these costs by assisting patients in numerous ways. Clinical health psychologists can implement interventions to improve knowledge and skill development for effective disease management. Psychologists are also prepared to assess and help patients modify maladaptive behaviors that may be negatively impacting upon health condition, as well as treating comorbid psychiatric problems that may also exacerbate the medical condition. As local clinical scientists, clinical health psychologists are uniquely trained to assess, implement, and evaluate the effectiveness of proposed interventions within disease management programs. In this study, the clinical health psychologist will work collaboratively with other interdisciplinary team members (e.g. physicians, nurses, registered dieticians, social workers, etc.) to create a developmentally appropriate education program on nutrition for adolescents with sickle cell disease. This training program includes education on the disease, education on the role of dietary intake in symptom management, and behavioral interventions aimed at improving dietary intake. However, a key component of this program also included the assessment of the adolescents' perceptions of their control over their lives while managing the

complications associated with sickle cell disease. Attention to attributional styles, including perception of control, self-efficacy and attitudes towards illness could help treatment teams identify the patients that may need more intensive psychological services to increase the likelihood of implementing suggested interventions.

Chapter Two: Literature Review

Background Information on Sickle Cell Disease and Traditional Treatment Recommendations

The term "sickle cell disease" refers to a collection of autosomal recessive genetic disorders that contain the Hb S variant on the hemoglobin gene (Ashley-Koch, Yang, & Olney, 2000). Approximately 80,000 Americans have sickle cell disease, making it one of the most prevalent genetic disorders in the United States (Ashley-Koch et al., 2000; Bonds, 2005; Smith, Oyeku, Homer & Zuckermann, 2006). Health care utilization reviews have reported an average of 75,000 hospitalizations per year among individuals with sickle cell disease (Ashley-Koch et al., 2000). In 1996, these hospitalizations cost over 475 million dollars per year (Ashley-Koch et al., 2000). Consequently, identifying mechanisms of more effective disease management that could reduce health care utilization costs is beneficial for individual patients, as well as for the larger society. In order to examine the role that nutrition can play in sickle cell disease management, it is essential to review some elements of the molecular pathogenesis of the disease.

The hemoglobin abnormality present with sickle cell disease affects oxygen absorption throughout the body, and when the red blood cell deoxygenates, the cell becomes stiff, sticky, and crescent shaped, like a sickle (Ballas, 2002). The polymerization process of the sickle hemoglobin plays a critical role in altering the structure and function of the cellular membrane of the red blood cells (Bunn, 1997). The rate and extent of polymer formation depends primarily on the intracellular hemoglobin concentration (Ballas, 2002; Bunn, 1997). Cellular dehydration occurs due to a loss of potassium and water, and is correlated with the polymerization process and increased

presence of irreversibly sickled cells (Ballas, 2002; Bunn, 1997). Although the exact mechanism by which sickle hemoglobin polymerization and cellular dehydration occurs remains unknown, traditional medical practice has always focused on the importance of hydration in disease management (Ballas, 2002). Changes in the cellular membrane that occur during red blood cell polymerization also increases its stickiness and the likelihood that the red blood cell will adhere to other cellular membranes (e.g., other blood cells or artery walls) and form barriers to the circulation of blood (Ballas, 2002; Bunn, 1997; Hagar & Vichinsky, 2008). The vaso-occlusive episodes are the physiological antecedents to acute pain, which derives from tissue damage secondary to the lack of oxygen (Ballas, 2002).

As a hematological disorder, the abnormalities in blood cell production affect organ systems and cause other common complications including increased rates of bacterial infection, chronic anemia, avascular necrosis, stroke, renal, and pulmonary disease (Cummin & Anin, 2003; Fung, Malinauskas, Kawchak, Koh, Zemel, Gropper, Ohene-Frempong, & Stallings, 2001; Ohnishi, Ohnishi, & Ogunmola, 2000). Although the focus of treatment for SCD is symptom management, current research is investigating ways of preventing vaso-occlusive crisis and subsequent organ and tissue damage (Ohnishi, Ohnishi, & Ogunmola, 2000).

Preventative treatments aimed at reducing the probability of vaso-occlusive crisis include transfusion therapy, hydroxyurea, and bone marrow transplantation (Claster & Vichinsky, 2003). In addition to the aforementioned microbiological features of a vaso-occlusive crisis, the red blood cells of patients with SCD also have a diminished life span of approximately 14 days, in comparison with 120 days, in people without this

hemoglobin abnormality (Ohnishi, Ohnishi, & Ogunmola, 2000). Transfusion therapy reduces this chronic anemia and increases the oxygen carrying capacity of blood cells by increasing the hemoglobin level and decreasing the percentage of sickled cells in patients (Claster & Vichinsky, 2003). Some patients with SCD are treated regularly with transfusions to decrease the probability of vaso-occlusive crisis; however, there are risks associated with this therapy. Repetitive transfusions causes iron overload when not carefully monitored and managed; iron overload can lead to endocrine failure, cirrhosis, and cardiomyopathy (Claster & Vichinsky, 2003). Another treatment alternative, hydroxyurea, is a chemotherapeutic drug that has been found to increase the levels of fetal hemoglobin. Fetal hemoglobin does not form the crescent shape under deoxygenation (Cummin & Anin, 2003; Ohnishi, Ohnishi, & Ogunmola, 2000). Consequently, hydroxyurea has been successful in reducing the frequency of vaso-occlusive crisis in some patients with SCD (Claster & Vichinsky, 2003). The toxicity of the drug, however, makes it difficult for many patients to tolerate this therapy on a long-term basis; there are also very serious side effects, including bone marrow suppression, associated with this drug therapy (Claster & Vichinsky, 2003; Ohnishi, Ohnishi, & Ogunmola, 2000). Lastly, bone marrow transplantation has a curative effect when successful. Unfortunately, this procedure is rarely applied due to the strict type match requirements and risk of death associated if rejection occurs (Ohnishi, Ohnishi, & Ogunmola, 2000). With the significant risks associated with the reviewed preventative treatments, current researchers are investigating less invasive treatment approaches that could reduce the frequency of vaso-occlusive crisis.

Role of Nutrition in the Management of Sickle Cell Disease

As advances in medical practices have expanded the life expectancies of individuals with chronic diseases, there has been a growing awareness of the role of nutritional factors in effective disease management. However, there is a paucity of literature examining the role of nutrition among children/adolescents with sickle cell disease. The literature that is available focuses primarily on deficits, as opposed to examining the restorative or preventative components of nutritional factors. In order to understand how average dietary intake impacts this population, contemporary researchers have chosen both qualitative and quantitative research designs.

Preliminary studies on nutritional status among children/adolescents with sickle cell disease examined anecdotal reports from parents. Ievers-Landis and colleagues (2001) utilized a semi-structured interview to assess thirty-seven caregivers of children with sickle cell disease on common parenting difficulties with this population. Common stressors that were reported included nutritional concerns, particularly concerns regarding amount of caloric intake and sufficient nutrients from varying food groups (Ievers-Landis, Brown, Drotar, Bunke, Lambert, & Walker, 2001).

In 2004, Mitchell and colleagues conducted another qualitative study that focused exclusively on nutritional status and mealtime behaviors of children with sickle cell disease. Data extrapolated from these focus groups (n=20) identified concerns regarding low body weight and a pattern in which caretakers attended more closely to hydration than to dietary intake. Most parents reported fluctuations in dietary intake that was influenced by factors of the disease process (e.g., lower intake during crisis episodes or hospitalizations). Parents reported minimal concern because their children often resumed

normal eating habits upon recovery. Parents also reported making compromises regarding dietary intake, in which hydration was valued above compliance with meals. Strengths that were noted included the effectiveness of education provided to parents on the role of hydration in the sickling process (Mitchell, Kawchak, Stark, Zemel, Ohene-Frempong, & Stallings, 2004). However, parents demonstrated a limited understanding about the role of other nutritional factors on disease management. Another critically important theme that came from the focus groups was an emphasis on increasing the child's responsibility for disease management. Parents reported that older children needed to become independent in self-care, including meal preparation and monitoring of sufficient intake. Consequently, investigators emphasize the importance of advancing parent education on nutritional risks associated with sickle cell disease and the development of behavioral nutrition intervention programs to prepare both parents and patients for more effective disease management (Mitchell et al., 2004).

Although height and weight deficits have been commonly reported about children with Sickle Cell Disease (Heyman, Katz, Hurst, Chiu, Ammann, Vichinsky, Gaffield, Castillo, Kleman, Thaler, & Lubin, 1985; Silva & Viana, 2002; Zemel, Kawchak, Ohene-Frempong, Schall, & Stallings, 2007), Gray and colleagues (1992) conducted the first study that quantitatively compared the nutritional status and patterns of dietary intake of children with sickle cell disease to healthy controls. Despite a very small sample size (n=28; only 9 patients with sickle cell disease), they found statistically significant differences in height, weight, and levels of both zinc and vitamin A (Gray, Bartlett, Kolsa, Marcuard, Holbrook, & Horner, 1992). In this sample, reviews of 3-day food records indicated normal intake levels in comparison with same-aged controls, but the

records also indicated an elevated basal metabolic rate for children with sickle cell disease (Gray et al., 1992). Therefore, investigators suggested that documented height and weight deficits, as well as zinc and vitamin A deficiencies, were due to higher metabolic demands, as opposed to inadequate dietary intake (Gray et al., 1992). They recommended the development of specific nutritional guidelines for this population to address the elevated metabolic rate and energy demands faced by this population (Gray et al., 1992).

Additional studies have been conducted to examine the nutritional status and dietary habits of infants, children, and adolescents with sickle cell disease. During acute hospitalizations of infants and young children with sickle cell disease (n=16), dietary intake was found to fluctuate between 35-53% of recommended daily allowances (Malinauskas, Gropper, Kawchak, Zemel, Ohene-Frempong, & Stallings, 2000). Fung and colleagues (2001) collected repeated measures of intake and energy expenditure for patients with sickle cell disease (n=46) during hospitalization and three weeks following discharge. Again, they found poor dietary intake during acute illness, with most of the calories consumed coming from oral fluids (Fung et al., 2001). Three weeks post hospitalization assessments indicated improvements in dietary intake when compared to hospitalization rates, but intake remained lower than recommended daily allowances for healthy children (Fung et al., 2001). They also found that this pattern of insufficient dietary intake seemed to worsen as subjects approached late adolescence.

In 2007, Kawchak and colleagues reported findings of a longitudinal study (n=97) examining the dietary intake of children and adolescents with sickle cell disease. Dietary intake was measured via 24-hour recall during annual clinic appointments, over a four

year period. Study results demonstrated a decline in weight and body mass index over time. Dietary intake of vitamin D, vitamin E, folate, calcium, and fiber was poor for the entire sample, with 63-85% of the children falling below the recommended daily allowances (Kawchuk, Schall, Zemel, Ohene-Frempong, & Stallings, 2007). Age trends highlight a decline in protein and macronutrients as the children enter adolescence. Consequently, investigators suggested that the observed growth deficiencies become more pronounced in adolescence because the patients demonstrated reduced dietary intake during a critical period with heightened energy demands (Kawchuk et al., 2007). They recommended the development of nutritional training programs for adolescents with sickle cell disease, using behavioral interventions to focus on nutrition education and meal planning, to ameliorate the decline in nutritional status (Kawchuk et al., 2007).

Most of the early studies on sickle cell disease highlight growth deficits, with children demonstrating delayed skeletal and sexual maturation (Phebus, Gloninger, & Maciak, 1984; Platt, Rosenstock, & Espeland, 1984; Stevens, Maude, Cupidore, Jackson, Hayes, & Serjeant, 1986). The pattern of low growth percentiles typically becomes apparent in children between two and five years of age, and is more significantly pronounced in male children with HbSS Disease (Zemel et al., 2007). A comprehensive study of sickle cell disease collected data from multiple treatment centers on 2000 patients and found growth failures to be unrelated to socioeconomic status (Platt, Rosenstock, & Espeland, 1984). Since many children with sickle cell disease have normal height and weight at birth (Silva & Vianna, 2002), it is hypothesized that nutritional deficiencies contribute largely to the deficits observed later in life (Zemel et al., 2007).

Deficiencies in a variety of macronutrients and micronutrients have been linked to specific clinical features of sickle cell disease (Reed, Redding-Lallinger, & Orringer, 1987). Macronutrients such as protein and overall caloric intake are essential to growth, sexual maturation, and immune system functioning (Reed, Redding-Lallinger, & Orringer, 1987). Malnutrition has been associated with deficits in T-cell functioning, including a decreased number of T lymphocytes (Reed, Redding-Lallinger, & Orringer, 1987). Studies examining immune system functioning among individuals with sickle cell disease has highlighted decreased numbers of T-cells and T-helper cells, hypothesizing that these deficits contribute to the frequency and susceptibility for bacterial infections (e.g., pneumonia) among this population (Hernandez, Cruz, Santos, & Ballestar, 1980).

Zinc is another key macronutrient that is associated with growth, sexual development and immune system functioning (Prasad, 2002; Reed, Redding-Lallinger, & Orringer, 1987). Although the exact mechanism of zinc deficiency among individuals with sickle cell disease remains unknown (Zemel, Kawchak, Fung, Ohene-Frempong, & Stallings, 2002), studies reviewing the effectiveness of zinc supplementation have demonstrated some promising findings. Among adults with sickle cell disease, zinc supplementation has demonstrated improved immune system functioning, including increased T-cell and T-helper cell production, and reduced numbers of irreversibly sickled cells (Prasad, 2002; Brewer, Brewer, & Prasad, 1977). More recently, a longitudinal study examined the impact of zinc supplementation on children with sickle cell disease (n=42) and found zinc supplementation to be associated with greater linear growth (Zemel et al., 2002). However, it is important to note that immune system functioning was not assessed in this study.

Micronutrients that are important for individuals with sickle cell disease include folic acid, vitamin B, vitamin E, and vitamin A. Although it is commonly known that folic acid aids fetal development during pregnancy, it is also helpful to individuals with sickle cell disease for similar reasons. Healthy African-Americans have been found to have lower levels of red blood cell folate than the general population (Kennedy, Fung, Kawchak, Zemel, Ohene-Frempong, & Stallings, 2001). Folate plays a critical role in the development of red blood cells. Since individuals with sickle cell disease already have higher red blood cell turnover due to the polymerization process, daily folic acid supplementation has been a standard component of care for the past 50 years (Reed, Redding-Lallinger, & Orringer, 1987). Although recent research has found no statistically significant changes in growth or frequency of acute illness with folic acid supplementation (Kennedy et al., 2001), it remains beneficial for individuals with sickle cell disease via more indirect effects. Red blood cell folate and vitamin B_{12} deficiencies are associated with increased levels of homocysteine. Homocysteine is an amino acid that can damage the lining of cell walls, increasing the likelihood of cellular adherence and vaso-occlusive crises that could lead to stroke (Kennedy et al., 2001; Segal, Miller, Brereton, & Resar, 2004).

The role of vitamin B in sickle cell disease has shown its most promising effects through in vitro studies (Nelson et al., 2002). Numerous in vitro studies have demonstrated anti-sickling effects of vitamin B_6. Unfortunately, these findings have been difficult to replicate through in vivo studies (Nelson et al., 2002; Reed, Redding-Lallinger, & Orringer, 1987). To date, there have been no studies examining the effectiveness of vitamin B supplementation with children/adolescents with sickle cell

disease. However, studies on vitamin B_6 supplementation with adults have shown slight increases in hemoglobin and decreases the frequency of hospital admissions and emergency room visits (Natta, Kurantsin-Mills, & Garry, 1990).

Vitamin E has also been found to demonstrate anti-sickling effects (Kawchak et al., 2007; Ohnishi et al., 2000; Reed, Redding-Lallinger, & Orringer, 1987). During deoxygenation, the sickled red blood cell is susceptible to oxidative damage which decreases the lifetime of the cell and increases the likelihood of irreversibly sickled cells (Ballas, 2002; Bunn, 1997; Kawchak et al., 2007; Reed, Redding-Lallinger, & Orringer, 1987). In-vitro studies have demonstrated that the presence of vitamin E decreases the formation of irreversibly sickled cells (Ohnishi, Ohnishi, & Ogunmola, 2000; Reed, Redding-Lallinger, & Orringer, 1987). In-vivo studies with adults with sickle cell disease have demonstrated inconsistent findings, often due to small sample sizes and methodological variability. However, some studies have demonstrated a decrease in irreversibly sickled cells (Natta, Machlin, & Brin, 1980) and decreased number of painful crisis episodes (Ohnishi, Ohnishi, & Ogunmola, 2000) with vitamin E supplementation.

Vitamin A is essential for vision, and it also plays a vital role in the effectiveness of the immune system and epithelial walls of cells throughout the body (Schall, Zemel, Kawchak, Ohene-Frempong, & Stallings, 2004). Children with sickle cell disease often demonstrate vitamin A deficiency. Those patients that are deficient in Vitamin A also demonstrate increased frequency and duration of hospitalizations, with lower hemoglobin than children with sickle cell disease that have normal vitamin A status (Schall et al., 2004). Vitamin A supplementation has been shown to improve hematologic status and

immune system functioning, decreasing hospitalizations precipitated by vaso-occlusive episodes or infections (Schall et al., 2004).

While poor dietary intake is common among adolescents with sickle cell disease (Kawchuk et al., 2007), norms regarding eating habits are socially reinforced. Prior to identifying potential mechanisms of remediation, it is equally important to discuss the documented dietary habits of same aged peers in the general population. National surveys have consistently documented poor dietary habits of adolescents in the United States (Hoelscher, Evans, Parcel, & Ketder, 2002; Lytle, 2002; Munoz, Krebs-Smith, Ballard-Barbash, & Cleveland, 1997). The U.S. Department of Agriculture's Continuing Surveys on Food Intake assessed the number of youth meeting national recommendations for food group consumption (e.g., 5-8 servings of vegetables per day), and found that adolescents across ethnic and socio-economic groups reported consumption below the minimum recommendations in every food group (Munoz et al., 1997). When separated by age and gender, the average consumption for adolescent males (ages 12-19) did meet minimum recommendations for grains, vegetables and dairy; however, same aged females did not meet the average minimum recommendations for any food group (Munoz et al., 1997). Eighty-seven percent of the females and 71% of the males assessed in this survey (n=3,307) also consumed fewer than 2,200 calories per day, which is the minimal recommendation for average energy demands (Munoz et al., 1997).

The American Dietetic Association stresses that the intake of a wide variety of foods is preferred over the use of nutrient supplementation (American Dietetic Association, 1996). However, with numerous studies indicating that adolescents in the United States do not consume adequate dietary intake, some researchers have chosen to

assess the use and success of nutritional supplements (Stang, Story, Harnsack, & Neumark-Sztainer, 2000). In a study examining patterns of supplement use and dietary intake, 423 adolescents (ages13-18) were interviewed (Stang et al., 2000). Results indicated that 15.6% (n=66) reported daily use of supplements, primarily consisting of multivitamins without minerals (Stang et al., 2000). Although all of the adolescents had similar daily caloric intake, those that did not use supplements obtained a larger proportion of their energy from total and saturated fats rather than from carbohydrates (Stang et al., 2000). Despite the aid of nutritional supplements, many adolescents still consumed less than 75% of the recommended daily allowance (RDA) for several macro and micro nutrients. For the adolescents reporting daily supplement use, 51.5% (n=34) and 47% (n=41) were deficient in zinc and calcium respectively, and 36.4% (n=24) were deficient in Vitamins A and E (Stang et al., 2000). Among the adolescents reporting sporadic supplement use (n=77), deficiencies in zinc, calcium, and Vitamins A, E, and B_6 increased to 49.4%, 53.2%, 42.9%, and 31.2%, respectively (Stang et al., 2000). Finally, adolescents with no supplement use (n=280) received less that 75% of RDA of nutrients at alarming rates, including the fact that 63% of them were deficient in calcium; 55% were deficient in Vitamin A and E; 49% were deficient in zinc; and 37.5% were deficient in Vitamin B_6 (Stang et al., 2000). The authors highlight the need for primary prevention programs aimed at improving dietary intake among adolescents in the general population of the United States (Stang et al., 2000). With rising concerns about pediatric obesity there has been a gradual increase in nutritional studies for children, but most occur in school settings and focus on middle school children as opposed to adolescents (Hoelscher et al., 2002).

Review of salient literature has highlighted the need for targeted nutritional interventions for adolescents in general, with special attention to adolescents with chronic illnesses such as sickle cell disease. To date, however, these recommendations have neither been implemented nor documented, so the potential mechanisms of change remain unknown. Mechanisms of behavior change have been extensively researched in the field of psychology; within the cognitive-behavioral framework, it is argued that sustained behavioral changes are achieved with concurrent cognitive changes. Schema changes may include changes in one's view of self, view of others or changes in their view of the larger world or society. Prior to examining the specific components that should be included in a nutritional intervention for adolescents with sickle cell disease, it is necessary to review the critical cognitive processes that could influence behavioral change.

Impact of Attribution Styles and Cognitive Appraisals on Effective Disease Management

Theoretical Underpinnings: Attribution theory

An attribution is a psychological concept that explains how individuals interpret the causes of events in other people's behavior, and the causes of their own behaviors. In 1958, Fritz Heider first postulated theories regarding attributions, in which he theorized that people generally attribute the causes of events or behaviors in one of two ways (Aronson, Wilson, & Akert, 2004). He believed that people engaged in an inference process due to an inherent need to predict and control their environment (Fiske & Taylor, 1991). Internal or dispositional attributions make the inference that behaviors are attributed to internal characteristics such as attitude, motives or personality traits.

External or situational attributions infer that events or behaviors can be attributed to external factors such as social norms, acts of God, fate or chance. Although critical analysis of all behavior typically can identify both internal and external antecedents, Heider argued that most people have the tendency to assign one form or the other, with the preponderance towards utilizing more internal attributions when making inferences about the causes of behaviors in other people.

Darryl Bem chose to investigate how people infer their own reactions, emotions, and attitudes (Fiske & Taylor, 1991). He argued that people often look to environmental factors for clues to help them determine their internal reactions to events (Bem, 1972). His self-perception theory suggests that when people are assessing the motives of their own behaviors, they will first determine whether or not their behavior is under the control of external forces or from their own desires (Fiske & Taylor, 1991). Bem's theory on self-perception is pivotal to the development of attribution theory because his model is the first to suggest that individuals do not simply know, intrinsically, how they feel about all events and that affective responses are often influenced by perception.

Bernard Weiner built upon these early theories by developing a model of causal attributions for achievement (Fiske & Taylor, 1991). According to Weiner's model, locus (internal vs. external), stability (stable vs. unstable), and controllability will determine the emotional consequences of attributions (Fiske & Taylor, 1991). Critics of his model have argued that the controllability dimension is poorly defined and not applicable in all domains (Meyer, 1980). However, the importance of locus and stability dimensions has been consistently supported in the literature (Abramson, Seligman, & Teasdale, 1978; deJong, Koomen, & Mellenbergh, 1988; Vallerand & Richner, 1998).

Even though Weiner's model was based on studies utilizing scenario methodologies, studies examining causal attributions in real situations have also provided converging support for the validity of Weiner's model (Vallerand & Richner, 1988).

Locus of Control

While many psychologists were examining the application of operant conditioning principles to human behavior, Rotter formulated theoretical assumptions that were the building blocks for social learning theory. Rotter (1954) proposed that an individual's behavior is determined by the nature or significance of reinforcements within the environment and an individual's expectation that these reinforcements will occur. These expectations are strengthened or weakened by the reinforcements provided (or lack thereof) and are generalized only to the extent that one situation is perceived as similar to the original situation (Rotter, 1990). Therefore, an individual's ability to accurately assess and interpret the antecedents and causality of that reinforcement accurately will directly impact upon the likelihood of an individual repeating a particular behavior. From these principles of social learning, Rotter later developed his theory on the generalized expectations for internal versus external control of reinforcement (Carton & Nowicki, 1994).

Rotter defined internal versus external locus of control as "…the degree to which persons expect that a reinforcement or an outcome of their behavior is contingent upon their own behavior or personal characteristic versus the degree to which persons expect the reinforcement is a function of chance, luck or fate, is under the control of powerful others, or is simply unpredictable" (1990, p.489). A plethora of studies have been conducted to examine this construct and most have found that individuals with a high

internal locus of control are more likely to have superior cognitive processing abilities, better academic and vocational performance, better emotional adjustment, and better socialization skills than their external counterparts (Crandall & Crandall, 1983). However, many early researchers tended to dichotomize these worldviews; it is now more commonly accepted to conceptualize locus of control perceptions on a continuum (Nunn, 1998). It is a continuum in which individuals with a high internal locus of control styles are more likely to view success or failure as results of their own actions or efforts, and individuals with high external locus of control patterns are more likely to believe that outcomes are due to fate, luck or circumstances (Nunn, 1988). From a more multidimensional view of locus of control, it is probable that individuals may exhibit high internal beliefs in certain situations and high external beliefs in other situations (Wolf, Sklov, Hunter, & Berenson, 1982). Consequently, considerable effort has been made to develop measures that assess locus of control beliefs in specific life areas.

Although locus of control has been widely studied across multiple disciplines, there is little convergence in the results found (Fournier & Jeanrie, 2003). It has been postulated that the lack of convergence may be due to variability in the instruments used and variability in the manner in which investigators define the construct (Fournier & Jeanrie, 2003). Early theorists incorrectly defined locus of control as a stable personality dimension yet others have focused on the intrinsically positive qualities of internality and negative qualities of externality. Rotter addressed some misconceptions in later work (1975) when he focused on the importance of reinforcement value in the assessment of locus of control. He argued that when people assign a low value to the reinforcement, they may obtain high internality scores yet demonstrate passive attitudes or behaviors

typically associated with externality (Rotter, 1975). Rotter consistently reported that locus of control style influences a general response style while others have argued that it should be considered as a domain specific construct (Fournier & Jeanrie, 2003). The investigation of domain specific locus of control has led to the proliferation of more than thirty new scales in the past thirty years (Fournier & Jeanrie, 2003).

The impact of locus of control on health related behaviors continues to be extensively researched (Helgeson & Franzen, 1998; Jacobson, Hauser, Lavori, Wolfsdorf, Herksowitz, Milley, Bliss, Gelfand, Werlieb, & Stein, 1990; Kavanagh, Gooley, & Wilson, 1993; Mackenbach, Borsboom, Nusselder, Looman, & Schrijvers, 2001; O'Hea, Grothe, Bodenlos, Boudreaux, White, & Brantley, 2005; Sorlie & Sexton, 2004; Surgenor, Horn, Hudson, Lunt, & Tennent, 2000; Wallston, Wallston, Smith, & Dobbins, 1987; Wallston & Wallston, 1978). Early studies found that patients with high internal locus of control were more likely to seek and retain information about their medical conditions, more likely to adhere to their prescribed medication regimes and more likely to be consistent about making and keeping appointments with medical providers (Wallston & Wallston, 1978). However, subsequent research on medical adherence among varied populations of patients has rendered inconsistent findings. Some authors suggest that the proliferation of domain specific instruments (e.g., disease or tasks) has made it more difficult to compare results across populations and contributes to the inconsistent findings reported in many meta-analyses (Meijer, Sinnema, Bijstra, Mellenbergh, & Wolters, 2002; O'Hea et al., 2005).

In order to examine the impact of locus of control beliefs on disease management in a pediatric population, some researchers have chosen to investigate parental locus of

control styles. DeMaso and colleagues (1991) examined the impact of maternal locus of control style and parenting stress on the adjustment of children with congenital heart disease. The study assessed 99 mothers of children, aged 4-10 years, with congenital heart disease on factors that influence the child's adjustment to living with a chronic illness. The findings indicated that mothers with an internal locus of control style reported better adjusted children and less overall parenting stress (DeMaso, Campis, Wypij, Bertram, Lipshitz, & Freed, 1991). The authors hypothesized that mothers with an internal locus of control style probably took a more active role in their child's medical management, thereby supporting their child's adjustment (DeMaso et al., 1991). However, it is also possible that parents with high internal locus of control are more likely to perceive their child's behaviors as a direct reflection of their parenting efforts and less likely to report behavioral disturbances. The lack of variety (e.g., father, teachers, etc.) in behavioral reporters is another limitation to this study.

Bakarat and colleagues (2005) conducted a similar study examining the association of parental locus of control with quality of life for children with Sickle Cell Disease. Thirty-one primary caregivers of children (aged infant-11years) completed standard measures of parental locus of control, family functioning, and pediatric quality of life during an inpatient hospitalization for pain or infection. The authors found that the majority of the primary caregivers reported high external locus of control beliefs, and this belief pattern was negatively correlated with optimal family functioning (Bakarat, Lutz, Nicholaou, & Lash, 2005). However, the authors suggested that completing data collection during an inpatient hospitalization may have increased the likelihood that caregivers would endorse disruptions in family functioning and locus of control style

which involved beliefs about powerful others (e.g., physician), as having greater control over health and illness outcomes.

Since the burden of managing a chronic illness eventually transitions from primary caregivers to the identified patients, it is critical to understand and directly assess child and adolescent health beliefs. Early researchers (Campbell, 1978; Campbell, 1975; Lewis, Lewis, Lorimer, & Palmer, 1977) found that health beliefs reported by children often differed from their parents' beliefs or their parents' accounts of their children's beliefs, further substantiating the need for direct assessment. Literature on health locus of control, coping skills, or illness related adjustments pertaining to an adolescent population is scarce. However, a few studies (Burkhart & Rayens, 2005; Murphy, Thompson, & Morris, 1997) have examined the impact of cognitive appraisals on medical adherence among children and adolescents.

Murphy, Thompson, and Morris (1997) conducted a study examining the impact of health locus of control on diabetes adherence behaviors among adolescents between 12 to 18 years old. The study consisted of 40 adolescents with Type 1 insulin dependent diabetes; data was collected at an outpatient medical clinic. Participants completed a number of measures assessing cognitive appraisal styles, including the Piers-Harris Self Concept Scale (PHCSC), the Children's Attributional Style Questionnaire, the Children's Health Locus of Control Scale (CHLOC) and the Diabetes Health Locus of Control Scale. Medical adherence was measured by assessing the frequency of blood sugar checks by downloading the memory chips from the adolescents' glucometer. The authors found that adolescents with high external locus of control were less likely to check their blood sugar on a regular basis and also had higher hemoglobin A1c levels than their

internal locus of control counterparts. Although the study also identified the fact that demographic (age and ethnicity) and family factors (parental monitoring and exertion of control) contributed to 44% of the variance observed in health behaviors, they found that cognitive appraisals accounted for 32% of the variance produced within the study, with locus of control and attributional style accounting for 16% of that figure. This study highlights the multidimensional factors that influence adolescent adherence with medical treatment recommendations. The results demonstrated poorer compliance in older adolescents; however, decreased parental monitoring may also influence these findings. It was also interesting to note that cognitive appraisals about locus of control and self-esteem had greater correlation with adherence behaviors than the participants' appraisals about the degree of stress associated with diabetes management.

Burkhart and Rayens (2005) examined the relationship between self-concept, health locus of control and adherence to daily peak expiratory flow rate (PEFR) monitoring among children with asthma. The sample consisted of 42 children between the ages of 7 to 11 years. The data was collected during a 5-week asthma self-management training program at an outpatient clinic. Participants completed the Piers-Harris Self Concept Scale (PHCSC) and the Children's Health Locus of Control Scale (CHLOC) at the beginning and at the end of the intervention program. Adherence was measured at the end of the intervention program by recording the number of days that the children used the PEFR at least once. This study found adherence to be positively correlated with a high self-concept ($r=0.33$, $p=0.03$) and high internal locus of control ($r=0.30$, $p=0.05$). They also found that scores on the self-concept scale and the locus of control scale increased over the course of the intervention program, with the CHLOC

scores moving towards an internal locus of control by the end of the intervention program. Some strengths to this study include daily digital monitoring of the outcome measure (PEFR flows) in a minimally intrusive manner, as well as the incorporation of behavioral techniques in the disease education provided. The reported outcomes suggest that increased education about one's disease can increase the likelihood of adopting an internal health locus of control. However, some limitations include the potential bias in data collection because the measures were read aloud to the participants and verbal responses were recorded in the presence of their parents.

Self-Efficacy

While the locus of control construct addresses outcome expectations, self-efficacy is another key construct that is assessed when examining the role of cognitive appraisal's impact upon health-related behaviors. Self-efficacy refers to one's beliefs about his or her ability to perform specific behaviors required to attain a desired outcome (Bandura, 1977). Bandura's theories on self-efficacy differentiate between efficacy expectations and outcome expectations; efficacy is focused on self-perceptions but outcome is based on self, in relation to the environment (Gecas, 1989). Bandura (1986) postulates that self-efficacy beliefs can originate from various forms of information including actual experiences, vicarious experiences, verbal persuasion, and physiological states. Performance attainment, or the actual experience of success, is considered to have the most influential impact on self-efficacy beliefs because it provides the best evidence on an individual's ability to achieve the desired outcome (Bandura, 1986). Consequently, self-efficacy beliefs are not global judgments about one's abilities, but they are task-specific and subject to change over time (Bandura, 1986).

Criticisms of self-efficacy research include discussions about the universality of the construct in cultures in which mastery, self-reliance, and achievement are not similar to those in western society. Some authors have found race and ethnicity to affect self-efficacy and perceived control even when controlling for social class (Gecas, 1989). Lower self-efficacy among African-Americans in comparison to European Americans seems to be associated with history of racial discrimination and diminished access to resources (Gecas, 1989). Gender differences in self-efficacy research have also been reported with males demonstrating greater self-efficacy and mastery than females in our society (Gecas, 1989). Lastly, some suggest that the predictive validity of self-efficacy on task performance is contingent on the complexity of the identified task (Gist & Mitchell, 1992). Consequently, the diagnostic tools developed to measure self-efficacy should attend to the subordinate process required for the performance of complex tasks to enhance validity (Gist & Mitchell, 1992).

Self-efficacy is a construct that has been utilized to assess adjustment to chronic conditions including fibromyalgia, arthritis, diabetes, and cystic fibrosis (Barlow, Williams, & Wright, 1996; Bartholemew, Parcel, Swank, & Czyewski, 1993; Buckelew, Murray, Hewitt, Johnson, & Huyser, 1995; Griva, Meyers, & Newton, 2000). These studies found higher levels of self-efficacy to be associated with reduced physical symptomatology (e.g., severity of pain) and psychological functioning (e.g., lower levels of depression). In addition to current levels of physical and psychological symptomatology, there is some evidence to support the idea that self-efficacy can affect the course of sickle cell disease across time (Edwards, Telfair, Cecil, & Lenoci, 2001).

There is a lack of extensive research on the role of self-efficacy in sickle cell disease management, due in part to a fairly recent development of a psychometrically validated, disease-specific measure (Edwards, Telfair, Cecil & Lenoci, 2000). However, Edwards and colleagues (2001) utilized a longitudinal study to asses the role of self-efficacy in sickle cell disease adjustment among adults (n=147). Their study found that adults with sickle cell disease reporting lower levels of self-efficacy also reported more physical and psychological symptoms, and greater health-care utilization than their counterparts with higher self-efficacy (Edwards et al., 2001). They also found that self-efficacy reports remained stable over the one-year study period, but highlight the fact that their study did not offer any interventions to address self-efficacy or other coping skills to support adjustment (Edwards et al., 2001). Strengths of this study included the assessment of multiple physical symptoms associated with sickle cell disease (e.g., pain, fatigue, nausea, etc.) and incorporated the frequency of emergency department visits in their assessment of health care utilization. Although the Sickle Cell Disease Self-Efficacy Scale demonstrated convergent validity with other measures of self-esteem, mastery, and locus of control, this new measure offers more disease-specific assessment of the role that self-efficacy beliefs can play on physiological symptoms and healthcare utilization. However, the prospective design used in this longitudinal study can only identify correlations and more experimental research is required to propose a causal relationship between self-efficacy and adjustment to sickle cell disease.

In 2007, Clay and Telfair replicated the previous study with adolescents living with sickle cell disease (n=148). Self-efficacy was measured by the Sickle Cell Self-Efficacy Scale (Edwards et al., 2000) and was compared with reports of physical

symptoms, psychological symptoms, and self-reports of their active participation in health-care (e.g., drinking adequate fluids, taking medicine, etc.). Results indicated that adolescents with higher self-efficacy engaged in more independent self-care behaviors and reported lower levels of physical and psychological distress (Clay & Telfair, 2007). In their discussion of the findings, the investigators highlight the need for brief, cognitive-behavioral interventions to be implemented, so that the role of self-efficacy in sickle cell disease adjustment can be experimentally assessed (Clay & Telfair, 2007). They also highlight the distinct differences that present during transition to adult healthcare and the need for training on adult life skill issues that are neither emphasized nor required for competent care in pediatric or family-centered care settings (Clay & Telfair, 2007). The investigators successfully validated the construct and convergent validity of the disease specific measure among adolescents with sickle cell disease. More experimental research with the measure is needed, but continued correlations between low self-efficacy and higher reports of physical and psychological distress suggest that this measure could be a beneficial screening tool in clinical settings to quickly identify adolescents with sickle cell disease that are at risk for poor adjustment.

Attitudes

In addition to locus of control and self-efficacy, clinical research has also focused on the role of attitudes in examining adjustment to a chronic illness. Attitudes have been defined in many different ways, but at the core of most definitions is an emphasis on their evaluative functions (Fazio, 1990; Fiske & Taylor, 1991). Affective, cognitive or behavioral factors may influence the evaluation of the stimulus and the attitude that is developed (Fiske & Taylor, 1991). Although research on attitudes dates back to social

psychological work in the 1930s, it remains a challenging construct to investigate because attitudes cannot be observed directly or even reported candidly. They must be inferred from human behavior (Fiske & Taylor, 1991). As previously discussed, self-perception theory argues that in the face of ambiguity, people also infer their attitudes from their own behavior (Bem, 1972).

When early social psychologists investigated the mechanisms of attitude change, some hypothesized that attention to inconsistencies is required for change. Festinger's cognitive dissonance theory (1957) postulates the idea that inconsistent cognitions about an event, object, or behavior will create dissonance. Festinger defines dissonance as an uncomfortable and aversive state of arousal. He suggests that most people will change either their cognitions or behaviors to improve consistency and reduce discomfort. While the presentation of information that may create cognitive dissonance can motivate change in behaviors and/or beliefs, there is evidence to suggest that many people engage in selective learning by attending only to attitude consistent information (Fiske & Taylor, 1991).

Research on adaptation to chronic illness has also examined the impact of children's attitudes about having a chronic illness on their overall adjustment and ability to utilize healthy coping mechanisms. As previously discussed, children with chronic diseases are at risk for developing behavior problems, emotional problems and social withdrawal (Austin & Huberty, 1993; Williams, Holmbeck & Greenley, 2002). Children that believe that their disease makes them different or less worthy than their peers are more likely to become more socially withdrawn and engage in maladaptive coping mechanisms (Austin & Huberty, 1993). Negative attitudes towards illness have been

correlated with internalizing and externalizing behaviors, such as increased anxiety, social withdrawal or disruptive behaviors, in children and adolescents with asthma and epilepsy (Austin & Huberty, 1993; Heimlich, Westbrook, Austin, Cramer, & Devinsky, 2000).

In a more recent study on the impact of attitudes towards illness on chronic pain in children/adolescents with arthritis, LeBovidge and colleagues (2005) found that attitudes played a moderating role between illness-related stress and depressive symptoms. The study utilized a structured interview with 75 participants (ages 8-18) to assess the impact of illness-related stressors on functioning, and also incorporated self-report measures on attitudes towards illness, depressive symptoms, and anxiety (LeBovidge, Lavigne, & Miller, 2005). Participants with a more positive attitude towards their illnesses reported lower levels of anxiety and depressive symptoms (LeBovidge, Lavigne, & Miller, 2005). Although both groups endorsed some levels of affective distress related to their medical condition, the authors hypothesize that having positive attitudes towards their illnesses may function as buffers that supports resiliency and decreases the likelihood of impairments in functioning (LeBovidge, Lavigne, & Miller, 2005).

Locus of control beliefs, self-efficacy beliefs and attitudes about one's illness may influence one's beliefs about health outcomes and ultimately affect one's willingness to engage in behavior changing activities. Yet some of the reviewed literature has suggested that targeted interventions may produce changes in the aforementioned thinking patterns, in addition to behavioral changes. With clarity on the nutritional deficiencies faced by this population and the critical cognitive constructs that may

influence behavior change, it is necessary to review the essential components recommended for designing an effective nutritional intervention for adolescents.

Designing Disease Self-Management Programs

Even though adolescence is a key developmental stage when many health behaviors are formed, a limited number of research studies have focused on physical health interventions specifically targeting adolescents (Williams, Holmbeck, & Grennley, 2002). Within the available literature, adolescent health psychology focuses on interventions aimed at preventing the onset of disease (primary prevention) or the early identification and treatment of select health problems (e.g., sexually transmitted infections) before a significant disease process has developed (Williams, Holmbeck, & Greenley, 2002). Interventions aimed at tertiary prevention are more highly focused on the management of chronic illnesses and constitute an area of adolescent health psychology that remains under researched, considering the rising incidence of chronic illness in our population (Williams, Holmbeck, & Greenley, 2002). An understanding of the impact of chronic illness on the biopsychosocial development of adolescents is critical for the progression of effective interventions aimed at supporting successful disease management (Williams, Holbeck, Greenley, 2002).

Adolescents with chronic diseases are more likely to display adjustment problems with higher incidences of internalizing symptoms and lower self-esteem than their healthy counterparts (Lavigne & Faier-Routman, 1992; Thompson, Gustafson, & Gil, 1995). The decreased self-esteem may be associated with delays in social development and/or body image concerns secondary to delays in pubertal development that is often

present with many chronic diseases, including sickle cell disease (Williams, Holmbeck, & Greenley, 2002). Although there is limited evidence about the impact of various illnesses on cognitive development, adolescents with sickle cell disease are at increased risk for cerebral damage secondary to stroke. These delays may impair problem-solving skills and other executive functions (Noll, Stith, Gartstein, Ris, Grueneich, Vannatta, & Kalinyak, 2001). Whether attributed to cognitive deficits or to intentional manifestations of behavioral autonomy, declines in adherence with medical regimes are consistent findings in adolescents, across multiple disease types (Quittner, Espelage, Ievers-Landis, Slocum, Seidner, & Jacobsen, 2000).

Focus on disease self-management groups originated with work focused on the rehabilitation of chronically ill children (Lorig & Holman, 2003). Thomas Creer and colleagues examined the role of health education on improving the outcomes of children with chronic asthma (Creer, Renne, & Christiam, 1976). Their work was pivotal, because they were the first to emphasize the importance of focusing on the active role of the identified patient in the treatment of chronic diseases, even when the identified patients were minors (Creer, Renne & Christian, 1976).

Although the medical perspective of a chronic disease course often focuses on physiological changes, some theorists have suggested that patients with chronic diseases often have fluctuating perspectives on their illness that does not correlate directly with physiological changes (Lorig & Holman, 2003). Patterson (2001) suggests that individuals with chronic diseases sometimes have illness in their psychological foreground and sometimes wellness is in the foreground, and that the goal of self-management programs is to help patients maintain a more consistent perspective on

wellness (Lorig & Holman, 2003). Corbin and Strauss (1988) conducted a qualitative study on adults with chronic illness and identified three essential tasks in effective self-management programs: medical management, role management, and emotional management. Medical management of a condition may include tasks such as taking medications, adhering to a special diet, or monitoring important levels such as glucose or blood pressure (Lorig & Holman, 2003). Role management focuses on tasks that allow patients to maintain or create meaningful behaviors that support life roles; and emotional management focuses on helping patients build coping skills for the psychosocial sequaelae associated with their illness (Lorig & Holman, 2003).

Self-management programs are most effective when based on a theoretical model, most often social cognitive theory (Gallant, 2003; Guevara, Wolf, Grum, & Clark, 2003). Social cognitive theory postulates the idea that what people learn and how they behave is influenced by their observations and thoughts about observed events (Bandura, 1989). Consequently, the likelihood of one demonstrating a desired behavior (e.g., adherence with diet) is influenced by an individual's access to the behavior modeled in their environment and their beliefs about their own ability to emulate the behavior demonstrated (Bandura, 1989; Gallant, 2003). Self-efficacy is viewed as the underlying mechanism that influences positive outcomes in self-management programs, thus enhancing self-efficacy is a key objective for self-management skills to be attained and utilized (Lorig & Holman, 2003). As previously discussed, self-efficacy beliefs can be influenced by performance attainment, modeling, social persuasion, and interpretation of physiological states (Bandura, 1986). Self-management programs can enhance efficacy by utilizing specific action planning to support success in skills mastery, while modeling

and social persuasion are often intrinsic components of the group interventions (Lorig & Holman, 20003). Psychoeducation about multiple contributors to physiological symptoms can also help patients identify a broader range of coping mechanisms to reduce and/or best manage their symptoms (Lorig & Holman, 2003).

Effectiveness of Disease Self-Management Programs

Cooper and colleagues (2001) conducted a meta-analysis on patient education programs that utilized behavior modification for adult patients with diabetes, hypertension, and arthritis. They highlight the methodological variability in patient education programs and the absence of theoretical models guiding intervention development in many studies (Cooper, Booth, Fear, & Gil, 2001). However, their analysis did reveal that programs using social learning techniques or cognitive-behavioral therapies demonstrated the largest effects (Cooper et al., 2001). The authors recommend that future trials identify a theoretical model of behavior change and measure psychological constructs such as quality of life or self-efficacy, preferably utilizing disease-specific measures, in addition to physical outcome measures like biochemical levels or frequency of healthcare utilization (Cooper et al., 2001). As a cautionary note, another meta-analysis on self-management educational programs among adults with chronic diseases conducted in 2004, highlighted evidence of a publication bias, in which studies indicating more positive physical outcomes were more prevalent in the literature (Warsi, Wang, LaValley, Avom, & Solomon, 2004).

In 2003, Guervara and colleagues conducted a meta-analysis on the effectiveness of self-management programs for children and adolescents with asthma. Their analysis of 32 studies with a total of 3,706 participants found that self-management programs

demonstrated modest to moderate improvement on numerous outcome measures including lung function, self-efficacy, reduced school absenteeism, and reduced frequency of emergency department visits (Guevara et al., 2003). However, this analysis incorporated studies with varying methodologies, including individual, group and family based interventions (Guevara et al., 2003).

There have been numerous systematic reviews and analyses of the effectiveness of self-management programs for children and adolescents with diabetes (Hampson, Skinner, Hart, Storey, Gage, Foxcroft, Kmbler, Cradock & McEvilly, 2000; Hood & Nansel, 2007; Northam, Todd, & Cameron, 2005; Winkley, Ismail, Landau, & Esler, 2006). Again, they each emphasized the importance of utilizing a theoretical foundation, with the use of psychoeducation and cognitive-behavioral techniques being most efficacious (Hood & Nansel, 2007). The systematic review determined that behavioral interventions focused on psychological and physiological outcomes demonstrated small to moderate effects, and those focused solely on physiological (metabolic) outcomes showed weak but statistically significant evidence of improved health (Hood & Nansel, 2007; Winkley et al., 2006). In addition to focusing on theoretical foundations and cognitive-behavioral techniques, the authors recommend that future trials should include disease-specific interventions that focus on action planning and opportunities for performance attainment within the intervention (Hood & Nansel, 2007). It is also important that interventions are designed with some degree of flexibility and adaptability for relevant socio-cultural factors that may arise in different populations of patients served (Hood & Nansel, 2007).

More recently, Kahana, Drotar, and Frazier (2008) conducted a meta-analysis on psychological interventions designed to improve adherence across pediatric chronic health conditions. The analysis included studies on adherence with medication, dietary, and/or exercise regimes that utilized pre-post evaluations or randomized control trials. A medium effect size was found for studies that utilized behavioral interventions (mean *d*= 0.54, n=10) and studies that were multi-component with both educational and behavioral interventions (mean *d*=0.51, n=46). When examining effectiveness across types of adherence outcomes, interventions aimed at improving self-care behaviors showed the largest effect size (mean *d*= 0.52, n=19), and dietary changes also demonstrating moderate effect size (mean *d*=0.47, n=19). The analysis also uncovered the fact that targeted interventions for children and adolescents with cystic fibrosis demonstrated the largest effect size (mean *d*= 0.74, n=13) with adherence outcomes greater than the results for diabetes studies (mean *d*= 0.38, n=24) and asthma (mean *d*= 0.23, n=37). In the management of cystic fibrosis, healthy weight is highly correlated with lung function (Stark, Quitner, Powers, Opipari-Arrigan, Bean, Duggan, & Stallings, 2009), and many of the interventions analyzed in this review (Powers, Jones, Ferguson, Piazza-Wagner, Daines, & Action, 2005; Goldbeck & Babka, 2001; Stark Opipari, Spieth, Jelalian, Quittner, & Higgins, 2003) included interventions targeting improved dietary intake. Further evaluation of the nutritional interventions implemented with this population could greatly inform the development of a nutritional training program for children and adolescents with sickle cell disease.

Nutrition Focused Management Programs for Children and Adolescents

Cystic fibrosis is an inherited chronic pediatric condition that affects primarily the lungs and digestive system (Milla, 2007). Malnutrition is a common side effect associated with cystic fibrosis due to pancreatic insufficiency and a variety of gastrointestinal disorders (Leonard, Davis, Rosenstein, Zeitlin, Paranjape, Peder, Maynard, & Mogayzel, 2010). Better nutritional status is associated with improved lung function, yet problems with dietary adherence and mealtime behaviors are often reported in this population (Leonard et al., 2010; Stark et al., 2009). With increased energy demands, similar to heightened need in sickle cell disease, it is recommended that patients with cystic fibrosis consume 120-150% of the recommended daily allowances for each essential food group (Stark et al., 2009).

Stark and colleagues (2009) designed a randomized control trial (n=67) to compare the effectiveness of a nutritional education intervention versus a behavioral and educational intervention on caloric intake and weight gain among youth with cystic fibrosis. Participants in both conditions attended 7 group sessions and were followed quarterly for 24 months after treatment. The parents and children attended separate manualized treatment groups over the course of 9 weeks. In the nutritional education cohort, the parents received training on how to measure the caloric content of meals, how to make weekly goals on increasing caloric intake, and how to incorporate higher caloric foods into their child's daily menu (Stark et al., 2009). The group of child participants (ages 4-12) used interactive games to increase knowledge about high energy foods and education on daily recommended dietary intake (120-150% of RDA) but they were not required to meet or rewarded for meeting these dietary goals. The behavioral plus cohort

followed the Be In CHARGE (Behavioral Intervention for Change Around Growth and Energy) protocol, in which parents were trained in behavior modification strategies in addition to nutritional information (Stark et al., 2009). Parents were instructed to create token economies and develop contracts in which their children could earn privileges for meeting caloric intake goals. Practice meals were presented in the behavioral plus children groups, and modeling and differential reinforcement was used to support increased intake of a wide variety of foods (Stark et al., 2009). Trophies were also given to the children if they met their caloric intake goals in the preceding week (Stark et al., 2009). Caloric intake data was recorded by parents on a daily basis and submitted to the treatment team during the parent sessions (Stark et al. 2009).

Children in the behavioral plus cohort demonstrated a larger increase ($p<.001$) in daily caloric intake, with an average increase of 872 calories per day, in comparison with an average increase of 489 calories per day in the education only cohort (Stark et al., 2009). The children in the behavioral plus cohort also demonstrated a larger increase in daily recommended allowances ($p<.001$), with an average increase to 148%, compared with an average increase to 127% among the education only children (Stark et al., 2009). However, the follow up data showed that the behavioral plus group made greater short-term improvements in caloric intake; however, over the long-term (24 months) both groups were performing similarly, consuming an average of 120% of RDA (Stark et al., 2009).

Strengths of this study include individualized goal planning during the treatment phase and opportunities for skill mastery and social persuasion within the treatment groups. In the long-term, the education only group probably performed so well because

there were elements of behavior modification built into the program. Although they were not explicitly reinforced with rewards in the education only cohort, both the parents and children were engaging in self-monitoring of daily caloric intake and the parents were receiving weekly feedback and support with problem-solving during their parent group sessions. Progress with improving caloric intake was also reviewed collectively in group sessions, so social persuasion was also present in the education only group. Although this study focused heavily on parent training because the youth were much younger, it provides an excellent framework for designing a similar behavioral and educational intervention on nutrition for adolescents with sickle cell disease.

When examining the literature for nutritional interventions targeting medically compromised adolescents, behavioral programs for teens with hypertension also present as good resources. Cardiovascular disease remains the leading cause of death in the United States and with the rise in childhood obesity; the prevalence of primary hypertension has also increased among American youth (Burrowes, 2010). Often the early signs of pediatric hypertension manifest as high blood pressure (Burrowes, 2010; Couch, Saelens, Levin, Dart, Falciglia, & Daniels, 2008). Typically, dietary modifications are the primary treatment approaches recommended to ameliorate elevated blood pressure in a pediatric population (Burrowes, 2010).

Couch and colleagues (2008) conducted a study (n=57) assessing the efficacy of a behavioral nutrition intervention on adolescents with elevated blood pressure. In this study, they modified a program previously successful with adults, the Dietary Approaches to Stop Hypertension (DASH) Diet, and compared blood pressure and nutritional outcome measures with youth receiving the DASH intervention to youth

receiving standard care. The DASH diet emphasizes increased consumption of fruits, vegetables, and low fat dairy, while simultaneously decreasing fat and sodium intake (Couch et al., 2008). The adolescents in the DASH intervention group (n=29) received 60 minutes of counseling with a registered dietician and a DASH manual that outlined food serving recommendations, food lists and tips for incorporating DASH foods into their diet, food monitoring instructions and guidelines for goal setting, action planning and self-rewarding (Couch et al., 2008). Over the three-month monitoring period, the DASH participants received eight weekly and two bi-weekly phone calls from interventionists, as well as bi-weekly mailings. Adolescents in this group also received monetary incentives for meeting weekly DASH goals in addition to compensation received for completing post-treatment assessments (Couch et al., 2008). The adolescents in the control or routine care group also received a 60 minute counseling session with a registered dietician. However they received only a booklet on healthy eating habits commensurate with the Food Guide Pyramid and this resource did not include information on specific food serving recommendations or information on self-monitoring and goal setting skills to accomplish dietary changes. Youth in the control condition did not receive intermittent contact, via phone or mail, during the treatment period and received monetary incentives only for compliance with post-treatment data collection (Couch et al., 2008).

Post-treatment comparisons revealed that the DASH intervention group demonstrated a decrease in systolic blood pressure ($P<.01$) that was greater than the routine care group (Couch et al., 2008). In fact, 50% of the DASH intervention group achieved blood pressure normalization, compared with 36% of the control group, in spite

of the fact that the intervention group had higher initial blood pressure measures than the cohort receiving standard care (Couch et al., 2008). The participants in the DASH intervention group also demonstrated a greater increase in the intake of fruits ($P<.001$), potassium ($P<.001$), and magnesium ($P<.01$) (Couch et al., 2008). Although the DASH participants increase in vegetable intake did not meet similar statistical significance ($P<.1$), their vegetable consumption at post-treatment and follow-up was double the amount reported in baseline (Couch et al., 2008).

Although the treatment approach in this study was individualized, the participants in the DASH intervention received detailed education on dietary recommendations, which included specific lists and tips on how to add heart healthy foods into their diet. Self-monitoring was encouraged and behaviorally supported with monetary incentives for weekly goal attainment in changes to diet. Regular telephone contact with the interventionists provided a mechanism for social support and assistance with action planning, as well as some level of social persuasion since the weekly data collection could reinforce accountability with food monitoring. One limitation that the authors did acknowledge was difficulty with retention in dietary research with adolescents. Nonetheless, the study outcomes demonstrated that significant changes can be achieved when behavioral interventions supplement traditional dietary education.

Lastly, Hoelscher and colleagues (2002) conducted systematic review of nutritional interventions for adolescents in the general population, to identify the critical components of designing effective nutritional intervention programs. They concluded that attributes of successful programs included a behavioral focus, interventions based upon a theoretical framework, attention and remediation of environmental factors (e.g.,

informing parents about study objectives to support family efforts at changing food selection options), having an adequate amount of exposure (duration and frequency of sessions), incorporating peer involvement, and providing mechanisms for self-assessment and feedback (Hoelscher et al., 2002; Lytle, 1995). When developing nutritional interventions focused on performance, and not only knowledge outcomes, training must include focus on specific foods that are rich in desired nutrients as opposed to nutrient education alone (Hoelscher et al., 2002). Strategies and methods that are utilized to accomplish program objectives should be practical and disseminated in developmentally appropriate language (Hoelscher et al., 2002). For example, interventions focused on meal planning and cooking demonstrations should incorporate nutritious food that adolescents can reasonably be expected to prepare independently.

Conclusion

Sickle cell disease is a hereditary blood disorders affecting over 70,000 Americans. Individuals living with sickle cell disease cope with frequent disruptions in normal routines, often necessitating urgent or acute hospital care, due to pain or infection. While many individuals with sickle cell disease are living longer, there have been limited medical advances in preventing vaso-occlusive crisis or subsequently preventing the sequalae of medical complications associated with repetitive vaso-occlusive episodes such as organ damage, avascular necrosis or elevated risks for pulmonary disease.

Review of the literature on the role of nutrition in sickle cell disease management demonstrates that growth and nutritional deficiencies have been documented in this population for a long time, yet few interventions have been designed to address these nutritional concerns. Close examination reveals that early parent education on disease

management has disproportionately focused on hydration, even though most treatment teams acknowledge the elevated energy demands in individuals with sickle cell disease and the importance of caloric intake. The vital role of specific nutrients, such as Zinc and vitamins B, E, & A, in immune system functioning and other hemoglobin properties are not routinely communicated to individuals with sickle cell disease in standard care. Nutritional deficiencies are often heighted during adolescence in this population, suggesting that psychosocial changes associated with this stage of development, including decreased parental monitoring and increased peer influence, may directly impact upon dietary intake. Although the need for behavioral interventions focusing on nutrition with this population has been discussed in the literature, to date, no disease-specific intervention programs have been reported for adolescents with sickle cell disease.

The development of any intervention targeting long-term behavioral change must consider the impact of cognitive appraisals on behavioral change. Theories regarding attribution styles, self-efficacy beliefs, and attitudes have been reviewed, as well as a review of the literature on how these constructs have affected behavior change in disease management. Social cognitive theory is proposed as the most effective theoretical orientation to utilize when developing a self-management program. Consequently, an effective self-management program should include opportunities for modeling, social persuasion, and performance attainment, in addition to education about disease parameters, action planning, and behavioral recommendations.

The present study aimed to examine the feasibility and acceptability of a newly developed behavioral intervention. The intervention explored the potential treatment

effects of the psychoeducational group intervention on patterns of dietary intake, as well as knowledge about sickle cell and nutritional recommendations. The education focused on information about sickle cell disease, including information about the increased energy demands associated with sickle cell disease, and also training about how to enhance one's diet with the key macro and micro nutrients that may potentially reduce disease complications. While exploring knowledge and dietary outcomes, the manner in which health locus of control beliefs may influence behavioral activation and adherence with dietary recommendations was also examined. Finally, we assessed what effect, if any, that participation in a nutrition focused self-management program would have on the participants' self-efficacy beliefs and their overall attitude about having sickle cell disease.

Chapter Three: Hypothesis

Overall Question:

This pilot study examined the feasibility of a five-week group intervention focused on nutrition and sickle cell disease. The pilot study primarily assessed the feasibility and acceptability of the five week intervention. Study goals also included exploration of potential treatment effects by monitoring changes in knowledge and dietary habits of adolescents with sickle cell disease. The study also explored cognitive appraisal styles by assessing health locus of control styles and disease specific self-efficacy beliefs at multiple intervals. Lastly, the study explored the potential impact of the self-management training program on the adolescent participants' overall attitudes about living with sickle cell disease.

Hypothesis 1:

H_o: The five week nutrition focused self-management program will not be feasible with the target adolescent population living with sickle cell disease.

H_1: The five week nutrition focused self-management program will be feasible, as defined by a minimum of 66% of evaluable participants completing the five week intervention and post intervention activities.

Rationale: Patients and families served at the outpatient hematology clinic have been receptive to other psychoeducational programs offered in the form of teen and parent groups. Study design focused on offering adequate exposure to the content without being excessively time consuming or lengthy in the duration of each session or number of sessions. Daily contact with the lead investigator during the intervention

phase should also provide social persuasion to help maintain attention to study goals and potentially to enhance motivation and commitment to attendance.

Hypothesis 2:

H_o: The five week nutrition focused self-management program will not be acceptable to adolescent patients living with sickle cell disease.

H_2: The five week nutrition focused self-management program will be acceptable to adolescents with sickle cell disease, and to their parents, as defined by a minimum of 66% of evaluable participants' scores of 4 or greater on question three of the satisfaction survey.

Rationale: The five week self-management incorporated characteristics that have previously been successful in other nutrition and disease management interventions with children and adolescents. These critical components include the use of social cognitive theory in group design, adequate exposure to intervention content, peer involvement, and opportunities for self-assessment and feedback. It was hypothesized that developmentally appropriate activities such as games and hands-on demonstrations within the group sessions would enhance acceptability with the target adolescent population.

Hypothesis 3:

H_o: There will be no difference in knowledge about sickle cell disease after participating in the self-management training program.

H_3: Participants will demonstrate increased knowledge about sickle cell disease after participating in the self-management training program, as assessed by total score on the Transition Knowledge Questionnaire for Sickle Cell Disease pre and post intervention.

Rationale: After participation in the five week nutrition focused self-management program, it was anticipated that the priming and distributed practice of the didactic information relayed will lead to increased knowledge about sickle cell disease and general treatment recommendations.

Hypothesis 4:

H_o: There will be no difference in knowledge about nutritional recommendations for optimal disease management, as assessed by total score on the nutrition knowledge questionnaire, after participating in the self-management training program.

H_4: Participants will demonstrate increased knowledge about nutritional recommendations for optimal disease management after participating in the nutrition focused self-management program, as assessed by total score on the nutrition knowledge questionnaire pre and post intervention.

Rationale: Again, with distributed practice of nutritional education across five sessions, and the process of maintaining a daily nutritional intake log, it was anticipated that participants would be able to demonstrate increased knowledge about healthy eating habits and healthy food selection.

Hypothesis 5:

H_o: Participants will demonstrate no differences in the amount of servings from each food group consumed on a daily basis, as assessed by the 24-hour food frequency measure, after participating in the nutrition focused self-management program.

H_5: Participants will demonstrate increases in the number of servings consumed daily from each of the five essential food groups (dairy, fruit, vegetables, meat, and grains) after participating in the nutrition focused self-management program, as assessed by 24 hour food frequency logs collected pre and post intervention.

Rationale: Since the literature indicates that adolescents in the general population often consume less than 75% of the daily recommended allowance of nutrients, and that adolescents with sickle cell disease demonstrate equally deficient dietary patterns, it was hypothesized that baseline measures of each participant's daily food consumption would be less than 50% of the recommended daily allowance in each food group. The U.S. Department of Agriculture (USDA) currently recommends 6 ounces of grains per day which can be achieved in 7-11 servings per day. Two and a half cups of vegetables (3-5 servings) and 3 cups of fruits (2-4 servings) are also recommended daily. USDA recommends 3 cups (2-3 servings) of milk and/or dairy products per day, and 6 ounces (2-3 servings) of meat, poultry, fish or beans per day. It was hypothesized that participation in the disease-specific education on the role of nutrition and the use of behavioral interventions, including external prompts and reminder cues via text messages from group facilitator, meal planning activities, modeling and social persuasion, would lead participants to increasing their daily consumption of food from each food group. It was further hypothesized that changes

in dietary intake in the fruit, vegetable, and meat/protein groups would be attenuated because they contain foods that are rich in the essential nutrients reviewed including Vitamin A, E, B, folic acid and zinc. Consequently, it is expected that the adolescents' food frequency measures post intervention to reflect 75% or more of the recommended daily allowances for each of the food groups.

Hypothesis 6:

H_o: Participants locus of control styles were be assessed by the Children's Health Locus of Control Scale and participants with internal and external locus of control styles will demonstrate similar changes in daily food group consumption throughout the self-management training program.

H_{6a}: Participants with an external locus of control style will demonstrate no differences in the amount of servings from each food group consumed on a daily basis, as measured by daily food logs and post-treatment 24-hour food frequency form, after participating in the self-management training program.

H_{6b}: Participants with an internal locus of control style would demonstrate increases in the number of servings consumed daily from each of the five essential food groups (dairy, fruit, vegetables, meat, and grains), as measured by daily food logs and post-treatment 24-hour food frequency form, after participating in the self-management training program.

Rationale: Although it was anticipated that all participants will be able to demonstrate increased knowledge about nutrition and healthy eating habits, it was hypothesized that cognitive appraisal styles would impact upon one's ability to apply

knowledge gained via behavioral changes. It is hypothesized that participants with a high internal locus of control will be more likely to incorporate knowledge and engage in the activity planning required to implement healthy eating behaviors. As previously discussed, early literature on the impact of health locus of control and medical adherence has indicated that individuals with a more internal belief pattern to be more proactive in seeking information about their condition and following treatment recommendations to support optimal health. However, participants with a high external locus of control style may be more likely to believe that their eating habits are contingent on external circumstances outside of their control (e.g. finances, food accessibility) and they may also be less likely to believe that altering their eating habits would have a significant impact on their illnesses. With the high level of unpredictability surrounding crisis episodes within the course of sickle cell disease, it is highly probable that participants with an external locus of control style would be less likely to believe that they are capable of making behavioral changes that would improve their health.

Hypothesis 7:

H_o: There will be no change in the participants' reports of their self-efficacy beliefs, as assessed by total scores on the Sickle Cell Disease Self-Efficacy Scale, after participating in the self-management program.

H_{7a}: Participants with low self-efficacy at the onset of the study will demonstrate increased self-efficacy beliefs after participating in the self-management training program.

H_{7b}: Participants with high self-efficacy at the onset of the study will maintain high self-efficacy after participating in the self-management training program.

Rationale: The literature has suggested that participation in self-management programs can enhance self-efficacy by providing access to vicarious experience and verbal persuasion from peers that are engaging in proactive self-care activities. Opportunities for success within the group experience may also support improved self-efficacy and gradual changes in behavior overtime. Therefore, it was hypothesized that the participants with low self-efficacy at the onset of the study would report improved self-efficacy after participating in the self-management training program.

Hypothesis 8:

H_o: There will be no change in the participants' reports of their attitudes about their illnesses, as assessed by the weighted total scores on the Child Attitude Towards Illness Scale, after participating in the self-management program.

H_{8a}: Participants with negative attitudes about their illnesses at the onset of the study will report more positive attitudes about their illnesses after participating in the training program.

H_{8b}: Participants with positive attitudes about their illnesses at the onset of the study will maintain positive attitudes about their illnesses after participating in the training program.

Rationale: As previously discussed, participation in an effective self-management program will offer opportunities for enhancing self-efficacy by giving participants

information on ways that they can affect aspects of their illness, via re-interpretation of physiological states, and opportunities for skill mastery. Interaction with other peers with similar medical complications and more positive attitudes about their illnesses could also be an influential component of modeling. Consequently, it was hypothesized that participation in the self-management program will increase the likelihood of more positive feelings about their illnesses.

Chapter Four: Methodology

Overview

This single subject study was implemented to assess the feasibility and acceptability of a brief cognitive-behavioral intervention for adolescents with sickle cell disease. The pilot study explored potential treatment effects by monitoring for changes in knowledge and dietary habits following participation in the group. Cognitive outcomes were also explored by examining disease specific self-efficacy beliefs, health related locus on control beliefs, and attitudes about living with sickle cell disease. Each group session incorporated didactic training, interactive games, and modeling to support skill development for effective disease management.

Design and Design Justification

The feasibility study used a single subject study design on a cohort of adolescents living with sickle cell disease. The study incorporated a repeated measures design with pre-test, post-test, and 1 month follow-up measures following the five week intervention. Since the presence of sickle cell disease is not a variable that can be manipulated experimentally, a single subject design is best suited to assess feasibility and explore potential treatment effects of the nutrition training program on eating behaviors within this special population. A single subject design is particularly useful when assessing intervention effects. It can provide preliminary data to guide evidence based practice when randomized experimental group designs are not feasible for the target population.

Participants

Study participants were recruited from a local sickle cell disease center. Members of the hematology team helped identify patients that met the inclusion criteria. To be

included in the study the adolescent needed to be between 13-17 years old, have a hematological diagnosis within the sickle cell disease classification, including sickle cell anemia (Hb SS), sickle hemoglobin C disease (Hb SC), or sickle beta-thalassaemia (Hb Sβ), and be actively engaged in disease management at the clinic, as defined as completing an outpatient appointment with a hematologist within 60 days prior to the onset of data collection. Sickle cell patients with comorbid developmental delays or patients that lived farther than 25 miles from the hematology clinic were excluded from the recruitment sample. Recruitment flyers describing the study were mailed to the homes of 30 families identified by the hematology team; 15 families also received flyers during outpatient clinic appointments. Ten families contacted the lead investigator with interest in participating in the study and were scheduled for intake appointments to review informed consent, obtain adolescent assent, and collect pre-intervention measures. One family did not keep the intake appointment and was not responsive to attempts at re-scheduling, leaving 9 families enrolled in the study (Table 1).

Table 1 Participant Demographic Characteristics

ID	Gender	Age	Phenotype	Parent Participant(s)
01	Female	17	Hb SS	Guardian (Aunt)
02[a]	Male	15	Hb SC	Mother, Father
03[a]	Female	15	Hb SS	Mother
04[a]	Female	15	Hb SS	Mother
05	Male	14	Hb SS	Mother
06	Male	15	Hb SS	Mother
07[a]	Female	15	Hb SC	Mother, Father
08	Female	14	Hb SS	Mother, Father
09[a]	Male	16	Hb SC	Mother, Father

[a]**Active Participants**

There were five female adolescents and four male adolescents with sickle cell disease enrolled in the study; all were African-American. The average age was 15.11 years (*SD*=.92). Five of the enrolled participants were from single-parent households, with the remaining 4 participants living with both parents. The sample of enrolled

participants was mixed phenotypically with 44% of the adolescents having sickle cell anemia (Hb SS n=4), 44% having sickle hemoglobin c disease (Hb SC n=4), and one participant having sickle beta-thalassaemia. Data was collected on hospital utilization and 44% of the participants (n=4) reported hospitalization 1-3 times per year for pain or other sickle cell related complications. Thirty three percent of the participants (n=3) reported more frequent illness with, 3-6 hospital admissions per year. Only one participant (ID 05) reported regular admission, with an average of 6-9 hospitalizations per year. Individuals with sickle cell disease often manage painful episodes at home and 55% of the participants (n=5) reported having painful episodes 2-3 times per month. Thirty three percent of the participants (n=3) reported less frequent complications, with pain occurring less that once per month. One participant (ID 08) reported considerably more complications from experiencing painful episodes at least once per week.

Although nine families were enrolled to participate in the study, four of the families were lost to follow-up, prior to the first session of the group intervention. One parent reported that illness prevented her son from attending group and another parent's late term pregnancy and limited childcare options for siblings prevented her daughter from attending. The other two families were not responsive to contact from study team and the reasons for attrition remain unknown. In the remaining five families (Participants 02, 03, 04, 07, and 09) that actively participated in the intervention and follow-up phases, there were 2 male adolescents and 3 female adolescents with an average age of 15.2 (*SD*=.44). Two of the five remaining participants were from a single parent household, and the remaining three lived with both parents.

Measures

General Information Form. Information on relevant demographics, including age, gender, ethnicity, and household composition was obtained by requesting the parent or legal guardian to complete a General Information Form during the intake visit. Disease specific and health related information was also recorded on this self-report measure, collected from participant and guardian. The disease-specific information obtained included disease genotype, frequency of pain episodes, infections, and number of inpatient hospitalizations in the past year. Other health related information obtained included height and weight.

Transition Knowledge Questionnaire for Sickle Cell Disease. In order to assess disease awareness, the Children's Hospital of Philadelphia (CHOP) Transition Knowledge Questionnaire for Sickle Cell Disease was utilized. This is a 25-item, multiple choice questionnaire, developed to examine the etiology, clinical manifestations, and medical management of sickle cell disease (Newland, Cecil, & Fithian, 2000). It was developed for use specifically with adolescents and has been found to have good internal consistency (Cronbach $\alpha = 0.72$) with this population (Barakat, Simon, Lisa, & Radcliffe, 2008).

Nutrition Knowledge Questionnaire. A nutrition knowledge questionnaire was developed by the lead investigator to assess knowledge about basic nutritional information (e.g. food groups, recommended daily allowances of essential nutrients, etc.) and information about the role of macro and micronutrients that are important for individuals with sickle cell disease. The questionnaire consisted of 12 items, utilizing multiple choice, true or false, and open answer items.

24-Hour Food Frequency Form. A food frequency form was developed and utilized to gather information about each participant's dietary intake. This data was used to identify patterns of consumption, with attention to the number of servings from each food group and the amount of fluids consumed daily.

Sickle Cell Self-Efficacy Scale. To assess the potentially mediating effects of self-efficacy, the Sickle Cell Self-Efficacy Scale (SCSES) was also administered to the adolescents. It is a nine-item, 5-point Likert scale designed to examine the perceived ability of participation in daily functional activities, despite complications associated with sickle cell disease (Edwards, Telfair, Cecil, & Lenoci, 2001). Response categories range from *not at all sure* to *very sure;* higher scores indicate greater self-efficacy (Edwards et al., 2001). The SCSES has been found to have good internal consistency (Cronbach = .89) among adults with sickle cell disease (Edwards, Telfair, Cecil, & Lenoci, 2000). Although scores on the SCSES related positively to other measure of self-esteem, mastery, and internal locus of control, results also indicate that self-efficacy beliefs are often situation specific and not easily obtained in other more global personality measures (Edwards et al., 2000).

Children's Health Locus of Control Scale. The Children's Health Locus of Control Scale (CHLOC) is a self-report questionnaire with 20 statements about factors influencing health that the participants can either agree or disagree with (Parcel & Meyer, 1978). The 20 items are summed to obtain a total scale score, with higher scores (31 to 40) indicating an internal locus of control style and lower scores (20 to 30) indicating an external locus of control style (Parcel & Meyer, 1978). Three factors can also be assessed by this measure including 1) beliefs regarding internal control, 2) general beliefs

regarding the degree that health and illness are determined by luck or chance, and 3) beliefs about the degree that powerful others (e.g. physicians, nurses, and parents) determine health and illness outcomes (Parcel & Meyer, 1978). In prior research with children, this measure has demonstrated satisfactory levels of test-retest reliability (r = 0.62 to 0.80), internal consistency (r = 0.75), as well as construct and discriminant validity (O'Brien, Bush & Parcel, 1989).

Child Attitude Toward Illness Scale. The Child Attitude Toward Illness Scale (CATIS) is a self-report measure, administered to assess the participants' positive and negative feelings about having a chronic illness (Austin & Huberty, 1993). The measure is composed of thirteen, 5-point Likert scale items, with higher scores reflecting more positive attitudes and lower scores indicating more negative attitudes (Austin & Huberty, 1993). The CATIS has been found to have a good internal consistency (r= .77 to .89) and test-retest reliability (r= .77 to .80) with children and adolescents aged 8 to 17 years with epilepsy and asthma (Austin & Huberty, 1993; Heimlich et al., 2000).

Satisfaction Questionnaire. As a feasibility pilot study, data on participant satisfaction and qualitative feedback was also collected. A short-form feedback survey was developed for use at the close of each session. Participants were asked to rate how well they believe the session covered the designated objective using a 5-point Likert scale (1=Poor/Not At All; 5=Very Good). They also answered two open ended questions to offer their feedback on their favorite and least favorite components of the session. At the end of the intervention, adolescent participants completed a satisfaction survey that was developed to assess their opinion on the appropriateness and usefulness of the group intervention. Participants were also asked to comment on the practicality of using text

messages and micro blogging (Twitter) for data collection of daily eating patterns, as well as their overall evaluation of intervention goals and activities. During the one month follow-up home visit, parents were also asked to provide feedback on the appropriateness and usefulness of the intervention on a parent satisfaction survey.

Procedures

Data collection was mixed in this study with some measures (e.g., CHLOC, CATIS, SCSES, and knowledge questionnaires) collected before and after the intervention, while other measures were collected post intervention only (e.g., feedback and satisfaction surveys). Data on dietary intake was collected before and after the intervention by the lead investigator, as well as during the intervention via text messaging and micro blogging on Twitter.

Twitter is a free social networking and micro blogging service that enables its users to send and read messages known as *tweets*. Tweets are text-based posts of up to 140 characters displayed on the author's profile page and delivered to the author's subscribers who are known as *followers*. Senders can restrict delivery to those in their circle of friends or, by default, allow open access. Users can send and receive tweets via the Twitter website, Short Message Service (SMS) or external applications. The lead investigator created a restricted group on the micro blogging site Twitter, for each participant to use a de-identified, password protected, account to report their food intake. Participants were asked to maintain daily entries in their on-line food diary by providing a tally of food consumed. By using Twitter, the information reported by study participants was communicated to the investigator for review and submission to a study database. The study database recorded the daily number of servings from five essential

food groups for each participant. For example, if a study participant sends a message stating "I ate a bowl of cereal with milk and drank a glass of orange juice this morning," this was transferred to the study database as one serving of fruit, one serving of grain, and one-half serving of dairy. Twitter was used to minimize the burden of maintaining a written food diary because Twitter could be accessed via computer and handheld devices, making it more accessible and not susceptible to loss that may occur with pen and paper journals. During the course of the intervention, some of the adolescents reported difficulty accessing their study developed Twitter accounts. Consequently, only one active participant sent food log via Twitter; two of the active participants sent food logs via email, and two of the active participants sent food logs via text messages to the lead investigator. All active participants received daily reminders and positive reinforcement via the communication medium that they selected (e.g., Twitter, text, or email).

Good Nutrition 4 Me Living with SCD Program

The Good Nutrition program was a 5-week adolescent intervention, focused on improving the dietary habits of teens with sickle cell disease. During the five weeks, there were four adolescent group sessions and one family session. Each group session was held at the local outpatient clinic and lasted 1.5 hours. The family sessions were held individually, at each participant's home, and lasted for an hour. The group sessions were held in the evening and dinner was provided for the adolescent participants. Attention was given to food selection to ensure that dinners and snacks provided were models of balanced meals with foods rich in the nutrients that are important to people living with sickle cell disease. Table 2 provides a brief overview of the goals and objectives of each session.

The intervention groups were led by two adult facilitators, the lead investigator and a medical social worker for the sickle cell disease center. The lead investigator has a Masters degree in social work and was a doctorial candidate for a degree in clinical psychology. The lead investigator has extensive experience in facilitating psycho-educational groups and therapeutic groups in both medical and mental health settings. The medical social worker that co-facilitated the groups also has considerable experience in facilitating psycho-educational groups and support groups for parents and adolescents living with sickle cell disease. During study development, the lead investigator had consistent consultation with two pediatric dieticians for input on the nutritional education provided in the intervention.

Table 2
Overview of Group and Family Sessions in Good Nutrition 4 Me Living with SCD Program

Session	Goals	Objectives	Materials Used
Session 1	Build Alliance and Cohesiveness Increase Knowledge about SCD Orient to Self-Monitoring	Icebreaker Activities Developing Group Rules Didactic Training on SCD Intro to Twitter and Food Logs	Study Handouts on SCD Blank 24hr Food Frequency Form Videos and Props on Blood Cells Session 1 Feedback Form
Session 2	Attention to Daily Intake Reinforce Knowledge on SCD Increase Knowledge about general nutrition facts, important nutrients for SCD, and food sources for nutrients	Discussion on Food Logs Good Nutrition Game Didactic Training on Nutrition	Study Handouts on Nutrition 10 Tips to a Great Plate [a] Session 2 Feedback Form
Session 3	Attention to Daily Intake Reinforce Knowledge on SCD and Nutrition Develop meal planning and healthy food selection skills	Discussion on Food Logs Good Nutrition Game Didactic Training on Meal Planning	Study Handouts on Nutrition Labels and Kitchen Basics Study Menu Ideas and Recipe Book Session 3 Feedback Form
Session 4	Attention to Daily Intake Increase Parental Knowledge on SCD and Role of Nutrition Facilitate Parent-Teen Communication on disease management	Review of Food Logs Reverse Role Plays and Modeling to review Good Nutrition Workbook Information Meal Planning Activity	Study Handouts on Parent Guide to Promoting Healthy Eating and Meal Planning
Session 5	Attention to Daily Intake Reinforce Knowledge on SCD and Nutrition Performance Attainment in Meal Prep	Review of Food Logs Good Nutrition Game Meal Preparation Discussion on Impact of Self-Monitoring	Teen Participant Satisfaction Survey

[a] USDA Center for Nutrition Policy and Promotion (2011). DG Tip Sheet No. 1.

In the first session, the group started with icebreaker activities to assist in the development of therapeutic alliance and group cohesiveness. This session focused on general education about sickle cell disease. The goal of this session was to provide preliminary information on the increased energy demands present in sickle cell disease and the reasons why adequate dietary intake is an important component to disease management. PowerPoint slides and videos were used to support didactic education on sickle cell disease. Each teen participant received a Good Nutrition 4 Me Living with SCD workbook that included handouts summarizing the education provided on sickle cell disease. During this session, the lead investigator also reviewed the details of dietary intake self-monitoring (food diary via Twitter) with the adolescents and assigned each participant his or her de-identified study Twitter accounts. Prior to the close of the session, each adolescent was asked to complete a brief feedback form on the session. Transportation assistance (parking vouchers or tokens) was provided as needed.

The second session began with a short game activity (Good Nutrition 4 Me Living with SCD Game) reviewing the information provided in the previous session on sickle cell disease. The adolescent participants elected to join teams and the winning team earned prizes that supported nutritional goals (e.g., water bottles, cold snack carrying cases, on the go cereal bowls, etc.). Following the game, the lead investigator reviewed food diaries to begin the discussion on compliance and possible barriers to completion. During this session, 3-4 participants disclosed problems with accessing Twitter accounts and each participant identified an alternative communication mechanism (e.g., text or emails) for future communication and data collection. Education in this session focused on general nutrition facts about the food groups, the recommended daily allowances from

each food group, the amount of daily hydration recommended and the nutrients that have been found to be important for individuals with Sickle Cell Disease (e.g., folic acid, vitamin B_6, vitamin E, zinc, etc.). The groups also discussed how to identify foods that are recommended to meet these nutritional needs. Each teen participant received handouts summarizing the education provided on nutritional recommendations to add to his or her workbook. Again, PowerPoint presentations were used to support didactic education on nutrition and sickle cell disease. Prior to the end of the session, each adolescent was asked to complete a brief a feedback form on the session. At the end of Session 2 each adolescent participant received $15.00 as partial compensation for time and effort in study. Again, transportation assistance (parking vouchers or tokens) was provided as needed.

Session 3 began with homework review of food logs submitted in the previous week. Each adolescent participant was presented with charts summarizing his or her consumption and progress with dietary goals. The charts were offered as a mechanism for visual cuing and stimulated social persuasion as the teens compared their progress with peers during the discussion. Following the homework review, the participants played another round of the Good Nutrition Game to review the information provided in the previous session, and the winning team won small prizes as reinforcement. Education in this session focused on skill building relative to meal planning. Participants were given handouts to add to their workbooks on making use of food labels and healthy food selection for a balanced plate. Participants were also provided with a recipe book and guide to healthy menu ideas developed by the study to assist them in planning meals that are rich in the nutrients recommended for people with sickle cell disease. By the close of

session, each participant had an individualized menu for at least three meals they could prepare that would incorporate the nutrients important to sickle cell disease. The lead investigator also modeled meal planning skills by assisting the group with selecting the meal they wanted to prepare collectively in the final session. Prior to the end of the session, each adolescent was asked to complete a brief a feedback form on the session. Again, transportation assistance (parking vouchers or tokens) was provided as needed.

The fourth session consisted of individualized home visits to assess how effectively the adolescents had been able to transfer knowledge gained at the groups to the home setting. During this session the lead investigator used modeling and reverse role plays to aid the adolescents in reviewing salient information with their parents about increased energy demands in children and adolescents with sickle cell disease and the vital nutrients that can support immune system functioning. The family and lead investigator discussed progress over three weeks in monitoring and increasing daily food consumption. The adolescent participant was provided with another chart on daily intake during the family session, and the family and investigator could review the charts to identify patterns, areas of strength and areas in which each teen could use improvement. The team brainstormed collaboratively on ways that the family could support goals of increased healthy food intake and meal planning. The lead investigator modeled use of meal planning tools to identify ways that their son or daughter could become more independent in food preparation and healthy food selection. The team also discussed plans for implementation of homework assignment of completing at least 2 out 3 meals planned in preceding group session. Parents were provided with handouts on how to make meal planning a family affair and general guidelines on how to help their

teens engage in healthy food selection. Each family was provided monetary supplementation ($25.00 grocery store gift card) to alleviate financial constraints that may hinder compliance with meal preparation task.

The final group session included one last game activity to reinforce information taught over the preceding four weeks. Again, the winning team was able to select prizes for answering the questions correctly about sickle cell disease features, disease management recommendations, and nutritional recommendations. The remainder of the group session focused on group preparation of the selected meal and discussion about meal preparation at home, as well as final thoughts about information they had learned on the role of nutrition in disease management. Towards the end of the final group session, the group participants completed the post intervention measures, including the teen satisfaction survey. At the end of this session each adolescent received $15.00 compensation for time and effort in study. Again, transportation assistance (parking vouchers or tokens) was provided as needed.

One month following the last group session, home visits were conducted with each family to assess progress with dietary goals after external reinforcement was removed. Study measures were administered to adolescent participants for the final time and parent/guardians were also asked to provide their feedback on the project via the parent satisfaction survey.

Chapter Five: Results

Overview

As a feasibility study, this study was not adequately powered for efficacy analysis. Feasibility and acceptability were assessed by descriptive statistics on intervention completion and qualitative data on satisfaction obtained at the end of the study. To explore potential treatment effects, non-regression analyses were utilized. Standard mean difference equations have been found to be the most appropriate form of non-regression analysis to use when assessing intervention effects in single subject designs (Olive & Franco, 2008). Effect sizes were calculated for all outcomes by subtracting the pre intervention mean from the post intervention mean and dividing it by the pre intervention standard deviation. For the knowledge measures and the measures assessing cognitive appraisal styles, the pre and post intervention means reflect the average of the scores across the five active participants. The data on dietary intake was organized to reflect the percentage of recommended daily allowance for each food group, and again the mean reflects the group average percentage for each food group across the five active participants. Lastly, comparative analysis is provided on the pre intervention measures from the active and non-active (intake only) participants in the study.

Feasibility/Acceptability

Feasibility was defined as having a minimum of 66% of evaluable participants complete the five week intervention. Although the study sustained attrition between intake and the intervention phase, the five families that started the intervention completed the intervention and the one-month follow-up. Three of the five evaluable participants (60%) attended all five intervention sessions and all of the evaluable participants (100%)

attended four of the five intervention sessions. Subjects 02 and 04 missed one group session due to illness during the intervention. The information they missed was reviewed individually with Subjects 02 and 04 when they returned to group the following week. Five of the five evaluable participants also completed the one-month follow-up visit. Consequently, 100% of the evaluable participants completed the intervention. Acceptability was defined as having a minimum of 66% of the evaluable participants report overall satisfaction with a score of 4 or greater on the satisfaction surveys. Eighty percent of the adolescent participants reported overall satisfaction with the 5 week nutrition focused self-management program on their satisfaction surveys. One hundred percent of the parents reported overall satisfaction with the program.

Qualitative data was collected weekly from the adolescent participants during the group intervention. After the first group session focused on learning about sickle cell disease and getting acquainted with group members, the adolescent participants reported an average score of 4.25 on accomplishing agenda goals, based on a Likert scale ranging from 1-5. All of the teens reported enjoying learning about sickle cell disease and how the red blood cells work in their bodies. The feedback form used following the second group session asked the teens to indicate how well they believed the group accomplished the goals of learning about nutritional recommendations in general and specifically those related to sickle cell disease management. Again the participants had an average score of 4.75 on a Likert scale from 1-5. In session 2, all of the participants indicated that the jeopardy-like game was their favorite part of the session. The feedback following the third session was positive, with an average score of 4.6 on the Likert scale about accomplishing goal of learning how to plan nutritious meals. Again, all participants

endorsed the game as their favorite part of the session, and one participant added that she enjoyed the meal planning discussion on how to add in foods she does not normally eat. The adolescent participants completed their satisfaction surveys at the end of the fourth group session. All five of the evaluable participants reported that they believed it was a good idea to offer a teen group focused on nutrition. All five also reported that they found the information provided in the program to be very useful. The teens reported a range of 2 minutes to 15 minutes required to send messages about daily intake, and 2 out of the 5 evaluable participants found the process of sending food diaries to be "somewhat" time consuming. Two of the remaining participants reported that sending daily dietary intake was "a little" time consuming and one participant did not find the process of reporting intake time consuming at all. Four of the teens reported difficulty using Twitter to communicate daily food intake; however, they found the alternative option of sending text messages or emails "convenient" or "very convenient." Three of the five evaluable participants reported that they believed monitoring their daily food intake was useful. Four of the five participants indicated that it was either "useful" or "very useful" to report their dietary intake to the group leader, and three out of the five teens reported that they believed that they were in agreement with the group leader about the type of healthy eating goals they needed to set, rating this as "always" or "most of the time." Four of the teens reported that they would like to continue in the program if more sessions were available and that they would recommend the nutrition focused groups to other teens with sickle cell disease. When given the opportunity for open feedback on the satisfaction survey, two participants suggested that the intervention be longer in the

future with more group sessions, and one participant suggested using text messaging instead of Twitter in the future.

Parent feedback was collected during the one month follow up visit on the parent version of the satisfaction survey. All of the parents reported that they believed it was a good idea to offer a nutrition focused training program for adolescents with sickle cell disease. All of the parents also reported that they found the information provided in the intervention on nutrition and sickle cell disease to be either "useful" or "very useful." All of the parents reported that that they would be interested in continuing in the program if more sessions were available and that they would recommend the group to other parents of teenagers with sickle cell disease. The parents were also given an opportunity to provide open ended suggestions on improvements to the program; the suggestions offered included the following: more group sessions for longer time to monitor dietary intake and potential change; offering a similar program for parents and children at a younger age to work on their eating habits as soon as possible; sharing more information at general hematology appointments about nutrition and the role of nutrition in disease management; potential training for officials in school setting about healthy food options since many kids consume breakfast and lunch at school; and informing parents about nutrition needs early on (at diagnosis) so they can start pattern of healthy food selection early. Some parents also reported benefits of participating in the program beyond enhancing knowledge; these included increased communication with teens while practicing meal planning activities and finding the recipe book offered to be age appropriate and easy to follow so they could feel comfortable encouraging teens to cook independently.

Knowledge Outcomes

Each adolescent participant completed questionnaires to assess their knowledge about sickle cell disease and their knowledge about nutritional facts and recommendations on three separate occasions: pre intervention, post intervention and one month following the end of the intervention. Both the Sickle Cell Disease Transition Knowledge Questionnaire and the study developed Nutrition Knowledge Questionnaire have a maximum score of 100. Table 3 shows the group means and standard deviations across the treatment phases. Effect sizes were calculated by subtracting the pre intervention mean from the post intervention mean and dividing by the pre intervention standard deviation (Olive & Franco, 2008). As demonstrated in Table 3, the trend in effect size data on knowledge outcomes was moving towards improvement with minimal improvement on knowledge about sickle cell disease (d= 0.08) and moderate improvement on knowledge about nutrition recommendations on daily intake and nutrients vital to individuals with sickle cell disease (d= 0.73).

Table 3
Descriptive Statistics and Effect Size Estimates for Knowledge Outcomes Across 5 Cases

	Pre Intervention Mean	**Pre Intervention SD**	**Post Intervention Mean**	**Post Intervention SD**	**Effect Size**
SCD Knowledge	85.6	9.63	86.4	8.29	0.09
Nutrition Knowledge	49.14	12.63	58.31	14.73	0.73

Repeat measures on knowledge were collected one month following the end of the intervention to assess how well the group retained the information provided in the intervention after exposure and reinforcement activities were removed. Table 4 demonstrates the effect size estimates from pre intervention means and one month follow up means for knowledge outcomes. Knowledge about sickle cell disease remained consistent with a group average score of 85.6 (SD=8.29). Knowledge about nutritional

recommendations demonstrated trends towards continued improvement (*d*= 1.06) with an average score of 62.5 (SD= 14.73) one month following the intervention.

Table 4
Descriptive Statistics and Effect Size Estimates for Knowledge Outcomes: One Month Follow-Up

	Pre Intervention Mean	Pre Intervention SD	1mth Follow-up Mean	1 mth Follow-up SD	Effect Size
SCD Knowledge	85.6	9.63	85.6	5.36	0.00
Nutrition Knowledge	49.14	12.63	62.5	12.83	1.06

Dietary Outcomes

Data on food frequency was collected from each adolescent participant pre intervention, during the intervention, post intervention, and one month following the end of the intervention. The information collected was categorized according to the five essential food groups to assess how many servings each adolescent was consuming and whether or not they were meeting basic nutritional recommendations. With hydration being a very important component of sickle cell disease management, data on fluid intake was also collected. Prior to examining change across the treatment phases, the average amount of food consumption reported at pre intervention was compared with national averages on adolescent food consumption in Table 5.

Table 5
Comparing Adolescent Average Daily Intake: Population Means vs. Pre Intervention Means

Characteristics	N	Grains	Vegetables	Fruits	Dairy	Meat/Proteins
Recommendations		7-11 Servings	3-5 Servings	2-4 Servings	2-3 Servings	2-3 Servings
Population Mean[a], Males	618	7.3	3.4	1.1	2.4	2.3
Pre Intervention Mean, Males	4	4.5	1.5	2	1.25	2.5
Population Mean, Females	672	5.1	2.8	1.1	1.7	1.5
Pre Intervention Mean, Females	5	4.2	1.2	0.2	1	2.5

[a]Muñoz et al. (1997). Food Intakes of US Children and Adolescents Compared with Recommendations

Raw data on the number of servings consumed were converted to represent the percentage of recommended daily allowance (RDA) for each food group. Table 6 presents the group means and standard deviations of the percentages of RDA consumed across the treatment phases. Again, effect sizes were calculated by subtracting the pre intervention mean from the post intervention mean and dividing by the pre intervention

standard deviation (Olive & Franco, 2008). As demonstrated in Table 6 and Figures 1-Figure 5, the trend in effect size data on dietary outcomes were trending towards improvement. There was no change in meat/protein intake because all five of the cases were consuming the recommended 2-3 servings daily at baseline. Effects sizes across the remaining food groups ranged from 1.46 to 2.68, indicating large improvements in dietary intake immediately post intervention.

Table 6
Descriptive Statistics and Effect Size Estimates for Dietary Intake Across 5 Cases[a]

Food Groups	Pre Intervention Mean	Pre Intervention SD	Post Intervention Mean	Post Intervention SD	Effect Size
Grains	60	23.43	94.4	7.67	1.46
Vegetables	54.6	20.06	100	0	2.26
Fruits	30	44.72	100	0	1.57
Dairy	40	22.36	100	0	2.68
Meat/Proteins	100	0	100	0	n/a

[a] Group means represent Percentage of Recommended Daily Allowance for each food group

In order to assess the maintenance of behavior change after the external reinforcements were removed, data on dietary intake was also collected one month following the end of the intervention. Table 7 demonstrates that the trend in effect size maintained some improvement. The group maintained large trends towards improvement for grain and dairy consumption (*d*= 0.97 and 0.89 respectively), and moderate improvement in fruit consumption (*d*= 0.45). However, there was a slight decline in vegetable and meat/protein consumption one month following the intervention.

Table 7
Descriptive Statistics and Effect Size Estimates for Dietary Intake: One Month Follow-Up[a]

Food Groups	Pre Intervention Mean	Pre Intervention SD	1mth Follow-Up Mean	1mth Follow-Up SD	Effect Size
Grains	60	23.43	82.8	23.55	0.97
Vegetables	54.6	20.06	53	37.93	-0.08
Fruits	30	44.72	50	50	0.45
Dairy	40	22.36	60	22.36	0.89
Meat/Proteins	100	0	75.4	42.43	n/a

[a] Group means represent Percentage of Recommended Daily Allowance for each food group

On an individual level, Figure 1 shows Subject ID02's pattern of consumption across the treatment phases. ID02 consistently consumed 100% of RDA of grains, fruits, and meat/protein from baseline through follow-up. During the intervention, ID02 typically consumed 1 serving of vegetables daily (33% of RDA); however, at the end of the intervention he was consuming 3 servings (100% of RDA). One month following the end of the intervention, Subject ID02 reported consuming 2 servings of vegetables, which represents 66% of the RDA and a return to his baseline rate of consumption. During the intervention ID02 also typically consumed 1 serving of dairy daily (50% of RDA). By the end of the intervention he improved, consuming 100% of the RDA of dairy. One month following the end of the intervention ID02 reported consuming 1 serving of dairy (50% of RDA) which represents an improvement from his lack of dairy intake at baseline.

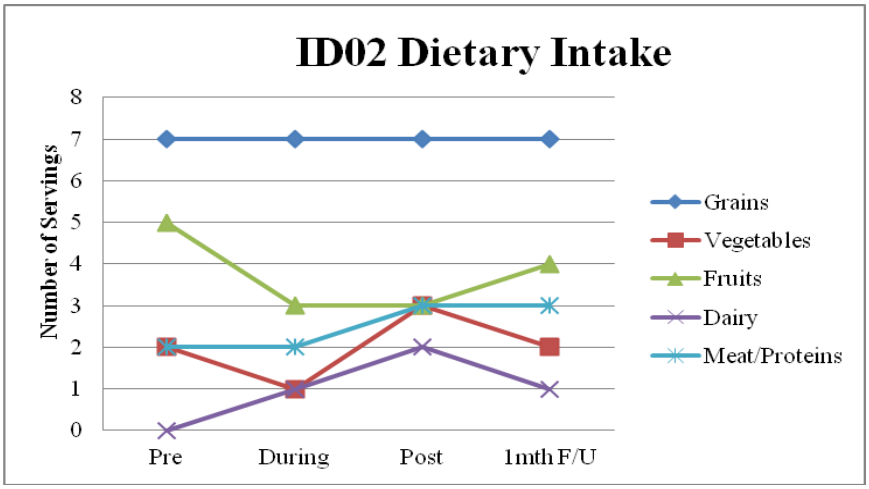

Figure 1 Dietary intake reported on 24hr Food Frequency Form across treatment phases. The number of servings presented "During" the intervention reflects the modal data on the amount reported over the 4 weeks of self-report.

Figure 2 shows Subject ID03's pattern of consumption across the treatment phases. ID03 consistently consumed 100% of the RDA of meat/protein across from

baseline through follow-up. At baseline, ID03 reported consuming 3 servings of grains (43% of RDA) and during the intervention ID04 typically consumed 4 servings of grains daily (57% of RDA). By the end of the intervention and during the follow up visit ID03 reported consuming 7 servings of grain (100% of RDA). During the intervention ID03 typically reported consuming 2 servings of vegetables daily (66% of RDA). At the end of the intervention, ID03 reported consuming 4 servings of vegetables (100% of RDA). However, one month following the end of the intervention she reported consuming 1 serving of vegetables daily (33% of RDA), which represents a return to her baseline rate of consumption. At baseline, ID03 was not consuming any fruit daily. During the intervention she typically reported consuming 1 serving of fruit daily (33% of RDA) and at the end of the intervention ID03 reported consuming 3 servings of fruit (100% of RDA). During follow up data collection, ID03 reported consuming 1 serving of fruit (33% of RDA). During the intervention ID03 typically did not consume dairy on a daily basis. At the end of the intervention she reported consuming 2.5 servings of dairy (100% of RDA), a rate of consumption she maintained at one month following the intervention.

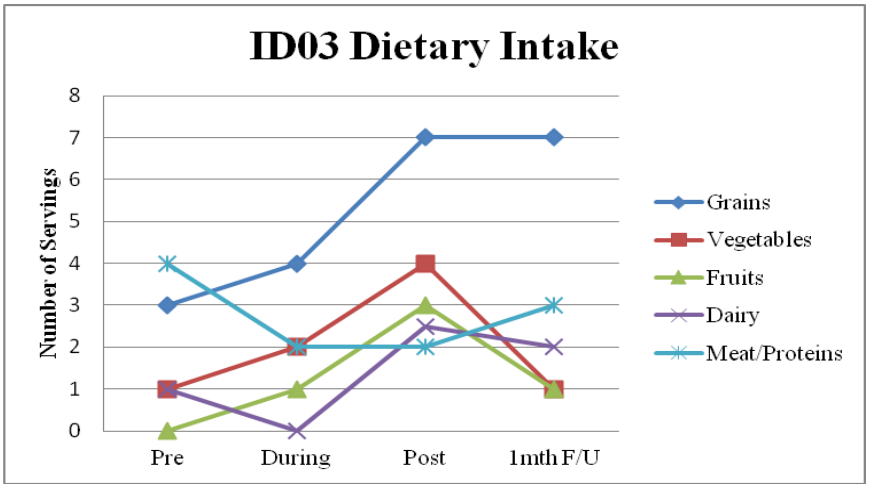

Figure 2 Dietary intake reported on 24hr Food Frequency Form across treatment phases. The number of servings presented "During" the intervention reflects the modal data on the amount reported over the 4 weeks of self-report.

Figure 3 shows Subject ID04's pattern of consumption across the treatment phases. During the intervention ID04 reported consuming 2 servings of grains most often (29% of RDA). At the end of the intervention she consumed 6 servings of grains (85% of RDA), and was able to maintain a rate of consumption of 4 servings of grains (57% of RDA) one month following the end of the intervention. At baseline, ID04 consumed 2 servings of vegetables daily (66% of RDA). However, during the intervention she typically reported consuming 1 serving of vegetable daily (33% of RDA). At the end of the intervention ID04 consumed 3 servings of vegetables (100% of RDA), yet during the one month follow up visit ID04 had not consumed any vegetables in 24 hours. At baseline and during the intervention, she typically did not report eating fruit. She reported having fruit only 9 out of 27 days of observation. During data collected at the end of the intervention ID04 reported consuming 3 servings of fruit (100% of RDA). One month following the intervention ID04 returned to her baseline rate of consumption with no fruit eaten in 24 hours. Prior to the onset of the study ID04 consumed 1 serving of dairy daily (50% of RDA). Most often ID04 did not report eating dairy during the intervention, and when she did report dairy consumption, it was typically one serving. At the end of the intervention ID04 consumed 2 servings of dairy (100% of RDA) and one month following the intervention she returned to her baseline rate of consumption of 1 serving of dairy daily (50% of RDA). At baseline, during, and post intervention ID04 consumed 3 servings of meat/protein daily (100% of RDA). During the intervention, most often ID04 ate 2 servings of meat/protein daily, which meet 100% of the recommended daily allowances. However, one month following the end of the

intervention ID04 had consumed only 1.5 servings of meat/protein in 24 hours (75% of RDA).

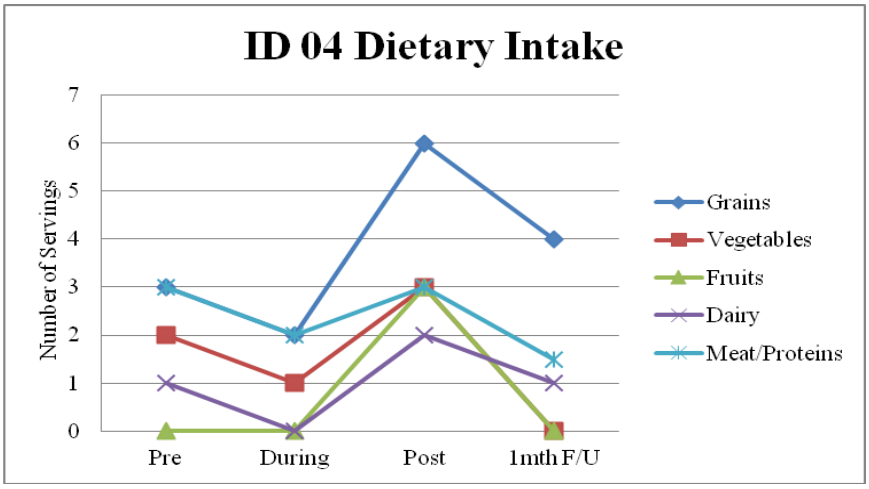

Figure 3 Dietary intake reported on 24hr Food Frequency Form across treatment phases. The number of servings presented "During" the intervention reflects the modal data on the amount reported over the 4 weeks of self-report.

Figure 4 shows Subject ID07's pattern of consumption across the treatment phases. ID07 consistently consumed 100% of the RDA of meat/protein across from baseline through follow-up. At baseline and during the intervention, ID07 typically consumed 4 servings of grains daily (57% of RDA). By the end of the intervention, ID07 reported consuming 6 servings of grains daily (86% of RDA) and she reported consuming 10 servings of grain (100% of RDA) at follow-up one month after the intervention. During the intervention, ID07 reported consuming 1 serving of vegetable most often (33% of RDA); however, she did demonstrate variability, consuming at least 75% of the RDA of vegetables 10 out of 20 days reported. At the end of the intervention, ID07 reported consuming 4 servings of vegetables (100% of RDA). One month following the end of the intervention she reported consuming 2 servings of vegetables daily (66% of RDA), which represents a return to her baseline rate of consumption. At

baseline, ID07 consumed 50% of the RDA for fruits. During the intervention, post intervention, and one month following the intervention, ID07 consumed 100% of the RDA of fruits. During the intervention, typically ID07 did not consume dairy; however, by the end of the intervention, ID07 reported consuming 4 servings of dairy (100% of RDA). One month following the end of the intervention, ID07 reported consuming 50% of the RDA for dairy, representing a return to her baseline rate of consumption.

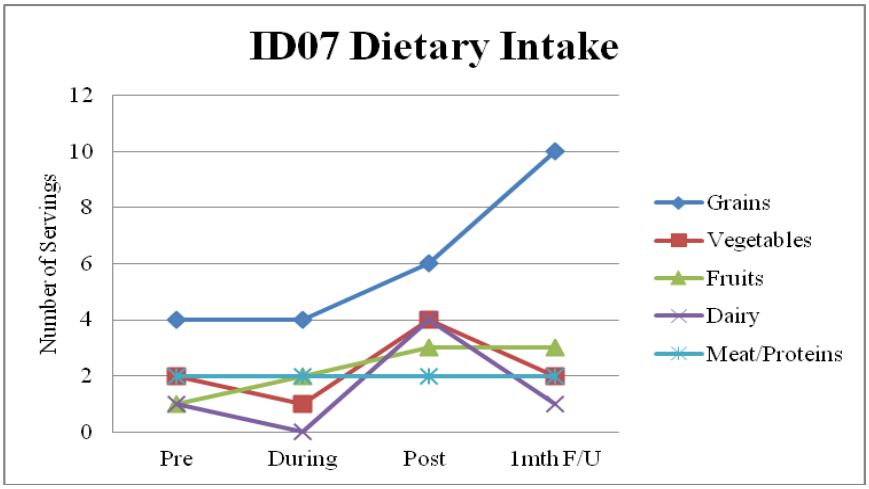

Figure 4 Dietary intake reported on 24hr Food Frequency Form across treatment phases. The number of servings presented "During" the intervention reflects the modal data on the amount reported over the 4 weeks of self-report.

Figure 5 shows Subject ID09's pattern of consumption across the treatment phases. ID09 consistently consumed 100% of the RDA for meat/proteins across the treatment phases. During the intervention ID09 typically consumed 5 servings of grains (71% of RDA), and by the end of the intervention he consumed 11 servings daily (100% of RDA). One month following the end of the intervention, ID09 reported consuming 4 servings of grains (57% of RDA). It is important to note that ID09 reported illness with joint pain for three days prior to the follow-up study visit and his parents reported a decline in dietary intake while ill. At baseline, ID09 reported consuming 1 serving of

vegetables daily (33% of RDA) and during the intervention, he typically consumed 2 servings of vegetables daily (66% of RDA). At the end of the intervention, ID09 consumed 3 servings of vegetables daily (100% of RDA) and he maintained this rate of consumption one month following the end of the intervention. At baseline, and most often during the intervention, ID09 did not consume any fruit. ID09 reported consuming fruit only 7 out of 29 days. However, post intervention ID09 reported consuming 4 servings of fruit (100% of RDA). One month following the intervention, he returned to baseline rates of consumption with no fruit intake. During the intervention ID09 also did not consume dairy regularly. Dairy intake was reported only 10 out of 29 days, with increased intake occurring after his family session in week 4 of the intervention. At the end of the intervention, ID09 reported consuming 3 servings of dairy (100% of RDA); however, at the one month follow-up visit, ID09 returned to baseline rates of consumption reporting one serving of dairy (50% of RDA).

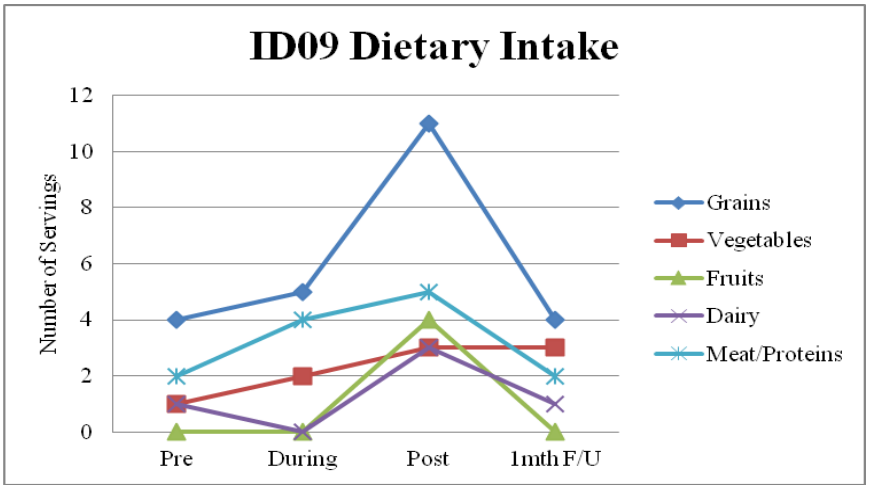

Figure 5 Dietary intake reported on 24hr Food Frequency Form across treatment phases. The number of servings presented "During" the intervention reflect the modal data on the amount reported over the 4 weeks of self-report.

Although hydration was not originally included in study hypotheses, maintaining adequate fluid intake is an important part of sickle cell disease management and daily

fluid intake was monitored throughout the study. The amount of recommended fluid intake varies according to weight, but the amount of fluid intake recommended for the adolescents that participated in the study ranged from 10 to 17 cups per day. Prior to the onset of the intervention, none of the evaluable participants were meeting these goals, with a group average of 5.4 cups of fluids daily. None of the evaluable participants were meeting minimal recommendations for fluid intake by the end of the intervention; however, the mean increased slightly to 6.1 cups daily. Effect sizes were calculated for hydration by subtracting the pre intervention mean from the post intervention mean and dividing it by the pre intervention standard deviation (SD=1.14) to reveal a moderate trend towards improvement with fluid intake (d= .61). Data collected one month following the end of the intervention showed a continuation of the trend towards improvement, with an average rate of fluid consumption of 7.7 cups daily (SD=1.3).

Cognitive Appraisals

The impact of cognitive appraisal styles were assessed by collecting responses to the Children's Health Locus of Control (CHLOC) questionnaire, the Sickle Cell Self-Efficacy Scale (SCSES), and the Child Attitude Towards Illness Scale (CATIS) prior to intervention, at the end of the intervention and 1 month following the end of the intervention. The CHLOC has a minimum score of 20 and a maximum score of 40, with lower scores (20-30) indicating an external locus of control style and higher scores (31-40) indicating an internal locus of control style. Scores on SCSES range from 9-45, with scores between 9-20 indicating low self-efficacy beliefs, scores from 21-32 indicating moderate self-efficacy beliefs, and scores from 33-45 indicating high self-efficacy beliefs. Lastly, the total weighted score of the CATIS ranged from 1 to 5, with higher

scores reflecting a more positive attitude about living with a chronic illness. Effect sizes were calculated and shown in Table 8.

Table 8
Descriptive Statistics and Effect Size Estimates for Outcomes on Cognitive Appraisal Styles Across 5 Cases

	Pre Intervention Mean	Pre Intervention SD	Post Intervention Mean	Post Intervention SD	Effect Size
SCSES	32.4	4.56	34.4	5.12	0.44
CHLOC	37.6	2.07	34.8	4.76	-1.35
CATIS	3.03	0.50	3.05	0.40	0.04

Changes in cognitive appraisal styles were mixed with a moderate trend towards improvement on sickle cell disease related self-efficacy beliefs (*d*= 0.44). However, there was a large effect trending towards regression on health locus of control beliefs (*d*= -1.35) and minimal effect (*d*= 0.04) on attitudes about living with sickle cell disease. At baseline, all of the evaluable participants' SCSES ranged from 29-37, reflecting moderate to high disease related self-efficacy beliefs. Scores improved gradually post intervention with the mean score of 34.4 continuing to reflect high disease related self-efficacy beliefs. One month following the end of the intervention two participants endorsed significantly lower self-efficacy beliefs, bring the mean score down to 30.2 (SD=5.93).

Prior to the onset of the intervention, all of the evaluable participants endorsed an internal locus of control style, with CHLOC scores ranging from 35-40 and an average score of 37.6. Although the scores on the CHLOC were slightly lower post intervention, 4 out of 5 evaluable participants continued to endorse an internal locus of control style. One month following the intervention 5 of 5 evaluable participants endorsed an internal locus of control style, with CHLOC scores ranging from 35-40 and an average score of 37.4.

Prior to the onset of the intervention, all of the evaluable participants reported both positive and negative emotions about living with sickle cell disease, with an average

CATIS score of 3.03. Following the intervention, the group continued to report mixed emotions about living with sickle cell disease, with an average score of 3.05.

Comparative Analysis Regarding Attrition

Four teens and their families were lost to follow-up after completing the pre intervention measures for the Good Nutrition 4 Me Program. Table 9 displays the mean scores and standard deviations on the knowledge and psychosocial measures for both the evaluable group (n=5) and the attrition group (n=4). The differences across the groups were not statistically significant; however, the teens that were lost to follow-up appeared to have less knowledge about sickle cell disease in general, and also seemed to be less confident about their abilities to effectively cope and manage the challenges they face related to sickle cell disease.

Table 9
Descriptive Statistics Pre Intervention: Evaluable Group vs. Attrition Group

	Evaluable Group Mean	**Evaluable Group SD**	**Attrition Group Mean**	**Attrition Group SD**
SCD Knowledge	85.6	9.63	67	18.58
Nutrition Knowledge	49.14	12.63	41.66	7.6
SCSES	32.4	4.56	25.5	2.66
CHLOC	37.6	2.07	35.75	2.63
CATIS	3.03	0.50	2.78	0.66

Chapter Six: Discussion

Summary and Interpretation of Findings

Children and adolescents with sickle cell disease are often at risk for nutrition and growth deficiencies (Heyman et al., 1985; Silva & Viana, 2002; Zemel et al., 2007). Prior research has shown that these deficiencies are often associated with a decline in dietary intake, a decline that is often exacerbated in adolescence (Kawchuk et al., 2007). This study presents the initial findings of a pilot project aimed as assessing the feasibility and acceptability of a newly developed behavioral intervention. The Good Nutrition program was focused on improving knowledge about the role of nutrition in disease management and improving dietary intake among adolescents with sickle cell disease. Outcomes show that the Good Nutrition program was feasible and acceptable to the target population. All of the parents indicated that an intervention focused on nutrition in sickle cell disease management was needed and useful for their families. All of the adolescent participants also found the information on healthy eating habits and important nutrients to be very useful. Most of the adolescent participants reported that the daily self-monitoring and communication with the group facilitator was equally beneficial. There were also high levels of overall satisfaction reported by the adolescents and their parents. Lastly, having 100% of the families that started the intervention remain and complete the intervention and follow-up data collection, is a testament to the palatability of the program. Although the outcomes are preliminary, they suggest there may be a strong interest in disease management programming focused on nutrition within the sickle cell disease population. The preliminary results also offer promising indications about the potential effectiveness of the intervention.

Potential treatment effects were explored in this study by assessing for changes in knowledge, dietary behaviors, and thoughts and feelings related to sickle cell disease. The changes in knowledge that were explored included knowledge about sickle cell disease and knowledge about nutrition. Nutritional knowledge focused on information about food groups and the number of servings recommended daily from each food group, information about the nutrients that are vital in managing sickle cell disease, and learning about food selection to accurately identify foods rich in the recommended nutrients. Study outcomes showed a significant improvement in knowledge about nutritional recommendations after participating in the Good Nutrition program. There were not observable changes in knowledge about sickle cell disease after participating in the program, but the participants were already well informed about sickle cell disease at the onset of the study. Close examination of the nutrition knowledge measures reflect a pattern of improvement in knowledge about daily recommendations in general, but more inconsistent awareness about the foods that are rich in the nutrients reviewed in the intervention.

Changes in dietary intake were also trending towards improvement after participating in the Good Nutrition program. Data collected immediately following the end of the intervention showed tremendous improvements with dietary intake goals. However, these gains were unduly influenced by characteristics of the study, with the post data collection occurring during the final group session, and study parameters including the provision of nutritious meals and snacks at every group. Consequently, it is important to focus on the potential treatment effects observed when comparing the dietary intake recorded pre-intervention and one-month following the end of the

intervention. Even after the positive reinforcements were removed, one-month following the end of the intervention, the participants showed improvements by increasing their consumption of grains, fruits, and dairy. The participants were already reporting adequate protein intake at the onset of the study, and this was maintained following the intervention. However, the participants did not demonstrate improvement in vegetable consumption.

As hypothesized, the adolescents with sickle cell disease were consuming less than 75% of the recommended number of servings of grains, vegetables, fruits and dairy prior to the study onset. In fact, the group was consuming less than 50% of the recommended number of servings of fruit and dairy at study onset. Review of the literature showed that adolescents in general, and female adolescents in particular, are consuming less that the recommended daily allowances from the five essential food groups (Muñoz et al., 1997). Findings from this feasibility study, as demonstrated in Table 5, show that the adolescents with sickle cell disease were consuming even less than their healthy counterparts across genders and across most food groups. Outcomes show promising indications about the potential treatment effects, with the participants consuming greater than 75% of the recommended number of servings of grains and protein, and 50-60% of the recommended number of servings of vegetables, fruit and dairy one month following the end of the program. Although changes in behavior related to fluid intake were not originally hypothesized, improvements in hydration rates were also observed after participation in the program. It is important to note that while the participants report of fluid intake increased throughout the study, the majority of the participants were only consuming 50-70% of the amount of fluids recommended daily;

highlighting an ongoing need for emphasis on both fluid and food intake in standard care for sickle cell disease.

Outcomes on changes in cognitive appraisals were mixed. Originally, it was hypothesized that locus of control style would influence behavioral changes, because prior research has found high internal locus of control styles to be correlated with greater adherence in disease management (Meijer et al., 2002; O'Hea, et al., 2005; Wallston & Wallston, 1978). This phenomenon was replicated in this feasibility study, with the participants endorsing high internal locus of control styles regarding health beliefs, and demonstrating improvements with dietary and fluid intake. However, with such a small sample, it is difficult to ascertain whether or not health locus of control beliefs are an important construct to assess with this population; and what mediating effects, if any, locus of control style has on behavioral activation and behavior change with this population.

The disease specific self-efficacy beliefs assessed in this study examined how well the adolescents believed they could manage their lives while living with the complications associated with sickle cell disease (e.g., pain, fatigue, etc.). Overall, the participants endorsed moderate self-efficacy beliefs, with most being uncertain about their ability to reduce pain and prevent symptoms from hindering enjoyment in activities. At the end of the intervention there was a trend towards improvement in self-efficacy beliefs, suggesting that the opportunities for performance attainment built into the intervention enhanced their beliefs about their abilities. However, one month following the end of the intervention, there was a trend towards regression, with self-efficacy beliefs reported being slightly lower than they were at baseline. Self-efficacy beliefs are

subject to change over time (Bandura, 1986). The decline observed in the data collected during the following up visits may be related to changes in physiological states. The follow-up data was collected at the beginning of the winter season and extreme temperatures can trigger complications in people with sickle cell disease. During the follow-up visits, all of the participants reported painful episodes in the month following the end of intervention, and three of the participants had been hospitalized for complications related to sickle cell disease. Comparatively, the data collected during enrollment was collected at the end of the summer and none of the evaluable participants had been hospitalized within 60 days of enrollment. The exposure to more negative aspects of sickle cell disease in the month following the end of the intervention most likely contributed to the slight decline in self-efficacy beliefs reported during follow-up.

At the onset of the study the adolescents reported mixed emotions about having sickle cell disease and this remained constant post intervention. All of the participants expressed frustration that this illness keeps them from doing activities that they like and hinders them from starting new activities, as well as causing them to feel different from other teens. Most reported feeling bad sometimes about having sickle cell disease. However, they also reported feeling happy, often, in spite of having sickle cell, not feeling as if having sickle cell was their fault, not feeling as if they would always be sick, and ultimately feeling like they are just as good as other kids. As observed in other studies, the participants' positive attitudes about their illness seemed to counterbalance the negative beliefs that were also reported, and supported resiliency (LeBovidge, Lavigne, & Miller, 2005). Although each participant had fluctuations on dietary intake

across the treatment phases, each participant's resiliency and positive functioning was observable in both group and individual trends towards improvement.

Relevance to Theory and Practice

The *Good Nutrition 4Me Living with SCD* program was designed to include established, efficacious techniques for behavior change including modeling, psychoeducation, action planning, and opportunities for social persuasion and performance attainment. These behavioral techniques have demonstrated success because they directly address the underpinnings of behavior change postulated by social cognitive theory; people will do what they observe others doing and what they believe they are also capable of accomplishing. Although the findings in this study have limited generalizability, it is evident that the behavioral interventions applied in this study were successful in supporting a trend towards improvement. Psychoeducation was moderately effective with the sample demonstrating improved knowledge about the role of nutrition in sickle cell disease management and general nutritional recommendations, after participating in the Good Nutrition program. Modeling and social persuasion also appeared to be influential, as the adolescents enjoyed the competition of the study developed game focused on sickle cell and nutrition knowledge, and they also frequently compared outcome charts on progress with dietary goals in the group sessions. Social persuasion and the participants' desires for positive reinforcement and praise were also evident in the family sessions, as progress with dietary goals were reviewed with their parents. Some of the participants also enjoyed the opportunities for performance attainment when completing meal preparation activities at home, and all participants were

afforded some opportunity for skill mastery during the joint meal preparation in the final session. With self-efficacy beliefs having an important impact on outcomes in self-management programs, the moderate, positive effect on sickle cell self-efficacy beliefs observed at the end of the intervention further support the benefit of the intervention program. The decline in self-efficacy beliefs reported during the follow-up visits appears to be associated with confounds outside of the control of the study, including change in season and change in disease severity during follow-up data collection.

While the primary aims of the Good Nutrition program were focused on enhancing knowledge and improving dietary habits, the study also sought to explore the benefits of using Twitter in a behavioral intervention. The use of various forms of mobile technology, including text messaging, video messaging, emailing, and traditional calling, has been explored with respect to monitoring adherence in disease management and appointment scheduling (Cole-Lewis & Kershaw, 2010). Although Twitter is a micro blogging, mobile tool, frequently used by teens and young adults in the social media age, its application in healthcare has not been explored or well documented in the literature to date. Results from this study indicate that Twitter may not be as beneficial or easily accessible as text messaging with the target population. Our findings were consistent with other behavioral interventions that used text messaging for communication and reinforcement in studies on diabetes management in adolescence (Franklin, Waller, Pagliari, & Greene, 2006). In future replications, the Good Nutrition 4 Me protocol should be changed to utilize text messaging as the primary tool of communication for reinforcement and data collection.

Outcomes from this study have important implications for clinical health psychology and the medical community managing sickle cell disease. As clinical health psychologists with a primary focus on health promotion, this study highlights the role that psychologists must play as scientist-practitioners in the healthcare system. As healthcare in general transitions towards more demands related to the management of chronic diseases, clinical health psychologists are uniquely prepared to develop and evaluate behavioral interventions focused on skill development for effective disease management. Although brief, the current feasibility study demonstrates the fact that there is interest and perceived usefulness in designing and implementing nutrition focused interventions for youth and families living with sickle cell disease. The program was developed from collaborative input from varied treatment providers, including hematologists, registered dieticians, nurses, and medical social workers. The program offered behavioral interventions focused on dietary intake that were previously recommended in the literature but had not been implemented to date. As local scientist-practitioners, clinical health psychologist can work with other medical professionals to develop single-subject design studies that will explore and potentially ameliorate emotional, cognitive, and behavioral barriers to effective disease management. Although single-subject design studies have their limitations, they are important because they perform a vital role in promoting better services for clients by supporting evidence based practice.

Small scale research by various members of the medical team is also beneficial because it can promote research activity amongst populations that are currently underserved. Disparity in research and clinical care in sickle cell disease has been documented for the past 40 years (Scott, 1970; Smith, Oyeku, Homer, & Zuckermann,

2006). Large discrepancies in public and private funding for research with sickle cell disease has persisted despite federal legislation aimed at enhancing parity (Smith et al., 2006). For example, in 2004, the prevalence of sickle cell disease in the United States was 2.66 times greater than the prevalence of cystic fibrosis, yet the National Institute of Health provided more funding for cystic fibrosis research and private funding outweighed sickle cell philanthropic support by more than 1.5 million dollars (Smith et al., 2006). While it is only speculative, "…we must consider the possibility that racial bias adversely affects the availability of resources, not only for research and the delivery of care, but also for the improvement of that care," (Smith et al., 2006, p.1767). As members of the medical team, clinical health psychologists have an ethical obligation to pursue equality in the provision of services for all patient populations. Psychologists can uphold these principles by engaging in research activities that support the development and implementation of clinical practice guidelines among populations that are marginalized and underserved.

Outcomes from this study also have implications for standard care in sickle cell treatment centers. This feasibility study substantiated earlier research findings, indicating that adolescents with sickle cell disease have inadequate dietary intake at baseline. The positive trends towards improvement with both knowledge measures and with dietary intake suggest that replication is warranted. Feedback from the families suggest that incorporating more education on red blood cell turnover, elevated energy demands, and nutritional recommendations is needed in standard parent training on sickle cell disease management. Lastly, certain aspects of the program, including electronic communication of dietary intake and resource information developed for the study, could be

advantageous for any treatment team member working individually with a sickle cell patient presenting with documented nutritional deficiencies.

Limitations of Current Study

As a preliminary feasibility study, there were multiple limitations to review. The attrition that occurred between enrollment and the intervention phase raises concerns about potential selection bias as a threat to internal validity. It is possible that the families that dropped out prior to the intervention phase had greater disease severity or less parental support, which may mean that disease severity and/or parental investment were moderators to the outcomes observed. Replication and expansion of the research design to include a control group would help address this potential threat to internal validity.

Potential threats to external validity were also present in this study including threats related to sample characteristics, reactivity, and threats related to the timing of measurements. In single-subject experiments it is crucial to collect as much information as possible about client characteristics; in order to address potential confounds to study outcomes. In this study, information about the family's socio-economic status was not collected, which could potentially be another confound to both the internal and external validity. Families with more financial resources may be better equipped to obtain healthy foods and accomplish eating goals, than other families from lower socio-economic groups. When replicating the study in the future, it will be important to collect information on socio-economic status.

Another potential confound and threat to external validity observed in this study is related to reactivity to the experimental arrangement. All of the participants were aware of the behavioral goals for dietary intake throughout the intervention, and it is possible that participants reported inaccurate dietary intake at the end of the study to please the investigator. In future replications, it would be advantageous to include a measure assessing social desirability (e.g., Marlow-Crown Social Desirability Scale or Children's Social Desirability Scale) in the post intervention data collection.

The timing of measurement can also influence validity and generalizability. In this study, post intervention data was collected during the last group session and consequently in a group setting. More errors were observed and addressed in this phase of data collection than during the baseline and follow-up data collections, which were done individually in the home setting. Information from the intervention (e.g., handouts, PowerPoint slides, etc.) were removed during data collection in the group setting but other features in the environment, including distractions from peers, could have influenced the post intervention outcomes. Meals were also provided at every group session; healthy meals that provided at least two servings from each food group. Therefore, as previously stated, the post intervention data on dietary intake collected during the final group session was unduly influenced by characteristics of the study environment. In future replications, it will be important to maintain consistency in data collection and have post intervention data be completed individually, ideally a few days after the last group session.

Future Directions

Preliminary outcomes of the *Good Nutrition 4Me Living with SCD* program suggest feasibility and acceptability with the target population. Replication is warranted to further assess the trends towards improvement observed. Future studies should consider the following changes to strengthen validity moving forward. Changes in enrollment plans are warranted to expand enrollment and minimize the impact of potential attrition. The groups should continue to be limited to 6-8 participants at a time, but different cohorts could be developed for access to a larger sample and data from a control group if a delayed treatment design is used. During enrollment, more information on socio-economic status and access to food resources should be collected. During enrollment, multiple collections of dietary intake should occur in order to have a minimum of 3 data points to average for baseline information on food consumption patterns.

During the intervention phase, future studies should consider expanding the intervention to 12 weeks, total duration. Literature on behaviorally based interventions for other disease populations often have a minimum duration of three months for proper dosage and for time to incorporate skills taught (Coles-Lewis & Kershaw, 2010). Expanding the duration could allow for multiple collections of pre intervention and post intervention data on dietary intake, for greater accuracy. It could also provide opportunity for an additional family visit to the home, since the family component appeared to have a significant impact on behaviors in this preliminary study. Results have shown that parental involvement appears to play an important role in knowledge application when it comes to food selection and meal preparation. Although the long-

term goals remain focused on enhancing knowledge and life skill development for effective disease management in adulthood, it is evident that the adolescents continue to need and respond to the guidance and support provided by their parents, even in the area of food selection and food consumption. In fact, providing simultaneous group meetings for the parents to receive education on sickle cell disease and nutritional recommendations would also be advantageous in future replications. Lastly, the additional time could afford the participants more time to focus on meal preparation and food selection, as it pertains to foods rich in the nutrients important in sickle cell disease management. Again, in future replications, post intervention data should be collected individually and include a measure on social desirability.

In addition to expanding the duration of the intervention, future replications should also incorporate more easily facilitated peer communication outside of the group sessions. Research among other disease populations has shown that peer support plays an integral role in disease management skill development, particularly skills utilized in social settings such as those related to food selection and dietary intake (Delamater, 2007; LaGreca, Auslander, Greco, Spetter, Fisher, & Santiago, 1995). Behaviorally based group interventions that incorporate facilitated peer communication have also shown promising effects in the diabetes management literature (Anderson, Wolf, Burkhart, Cornell, & Bacon, 1989; Greco, Pendley, McDonell, & Reeves, 2001). It would be advantageous to explore the potential benefits of enhanced peer support by adjusting the treatment protocol to include facilitated peer communication, via text messaging. The adolescents could be paired at the beginning of the intervention and be encouraged to communicate outside of group with their partner about progress with dietary goals.

Knowing now that the nutrition focused, behavioral intervention was beneficial, future studies should also focus on bringing uniformity to the procedures, to help guide clinical practice. The program should be standardized by developing a script for the study sessions and developing other quality assurance measures, including treatment fidelity checklists. Group sessions should be recorded to monitor for treatment integrity, and an independent rater should review at least 25% of the videos while completing the treatment fidelity checklists. The clinicians facilitating the study sessions should also complete fidelity checklists at the end of each session, and their reports should be compared with the checklists from the independent reviewers to assess for inter-rater reliability. Lastly, multiple raters should review the dietary intake reported and converted into servings across food groups, to again assess for inter-rater reliability.

Summary and Conclusions

In spite of the limitations reviewed, the *Good Nutrition 4Me Living with SCD* program appears to be a promising addition to standard care for adolescents with sickle cell disease. To date, this program is the first psychoeducational treatment study designed specifically to address nutritional concerns for adolescents with sickle cell disease. The program incorporated key components of behavior modification procedures including exposure, modeling, and positive reinforcement on a fixed interval. Opportunities for performance attainment and social persuasion in the group setting appeared to be beneficial in supporting positive change in knowledge and behaviors related to dietary intake. Hydration and dietary intake are important aspects of self-care in sickle cell disease management, and interventions focused on developing the skills associated with good self-care are an important component of comprehensive care.

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