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Health-related quality of life in children with sickle cell disease using the Child Health Questionnaire

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Abstract

Introduction—This study sought to determine if changes in parent-reported health-related quality of life (HRQOL) in children with sickle cell disease (SCD-SS) occurred after participation in a nutritional supplementation study, and to compare HRQOL responses with normative scores from non-Caucasian children.

Method—Parents of children with SCD-SS between the ages of 5 and 13 completed the Child Health Questionnaire (CHQ-PF50) at baseline and 12 months.

Results—For the 47 children (8.6 ± 2.4 yrs, 43% female), baseline CHQ scale scores were significantly lower than normative scale scores for parental emotional impact, general health, and overall physical health, but higher for mental health. After the nutritional supplementation study, overall physical health and parental emotional impact improved to normative levels. Furthermore, physical role functioning significantly improved.

Discussion—Participation in a nutritional study had a positive impact on parent-reported HRQOL physical scores in children with SCD-SS. More research is necessary to develop care providers' awareness and adequate HRQOL interventions for this population.

Keywords

Health-related quality of life; sickle cell disease; children

Introduction

Sickle cell disease is a chronic, inherited disease common in the African American population. The more severe form of sickle cell disease, type SS (SCD-SS), affects up to 1 in 400 African American children, and is associated with frequent pain, growth faltering,

sub-optimal nutritional and energy intake, organ system failure, strokes and death (Kawchak, Schall, Zemel, Ohene-Frempong, & Stallings, 2007; Tsaras, Owusu-Ansah, Boateng, & Amoateng-Adjepong, 2009). The World Health Organization (WHO) defines health as “a state of complete physical, mental and social well-being”. SCD-SS significantly impacts each of these domains (Thomas & Taylor, 2002). In addition to suboptimal physical well-being, children with SCD often have lower school attendance due to illness-related absenteeism, higher levels of emotional internalizing problems, lower levels of social competence and lower school performance compared with healthy peers (Trzepacz, Vannatta, Gerhardt, Ramey, & Noll, 2004). According to the Centers for Disease Control and Prevention, “health-related quality of life (HRQOL) is a multidimensional concept that usually includes self-reported measure of physical and mental health.” (<http://www.cdc.gov/hrqol/wellbeing.htm>). The Child Health Questionnaire (CHQ) is one of the best-known and widely used measures of HRQOL for children and adolescents (Landgraf, Abetz, & Ware, 1999; Raat, Bonsel, Essink-Bot, Landgraf, & Gemke, 2002). It has been useful in comparing health-related quality of life of children with chronic diseases and healthy controls (Drotar, 2004; Wallander, Schmitt, & Koot, 2001), and is a valid predictor of psychosocial and physical health (McCullough & Parkes, 2008).

Due to the complex nature of SCD-SS, HRQOL may be affected by both symptoms and treatment interventions (Palermo, Schwartz, Drotar, & McGowan, 2002). Children with SCD-SS are at an elevated risk for life threatening health problems compared with healthy peers (Quinn, Rogers, & Buchanan, 2004). Children with SCD-SS treated with chronic transfusion therapy report even greater problems with pain, school attendance, increased disease awareness and hospitalization (Stegenga, Ward-Smith, Hinds, Routhieaux, & Woods, 2004). Nonetheless, the current literature on HRQOL in SCD is sparse, and has focused primarily on disease severity. The few studies that have examined HRQOL in children with SCD indicate that those who experience more disease-related complications such as chronic transfusions, acute chest syndrome, asthma and pneumonia have poorer HRQOL, as measured by the CHQ (Palermo et al., 2002; J. A. Panepinto, O'Mahar, DeBaun, Loberiza, & Scott, 2005). Further, zinc supplementation in SCD is associated with potential markers of health related quality of life including reductions in painful crisis, decreased incidence of infections and accelerated growth (Brown, Peerson, Rivera, & Allen, 2002; Prasad et al., 1999). However, these results did not differentiate between SCD phenotypes; thus little is known about HRQOL in children with more severe forms of SCD disease. Currently, there is no descriptive information on the HRQOL of children with SCD-SS specifically, and no comparison with normative data about healthy children.

Understanding specific domains in which HRQOL is reduced in this population will help identify areas to target for future medical, psychological and behavioral interventions.

Access to more normative data on HRQOL in children with SCD-SS may allow for early detection of deficits, and early interventions (Anie, 2005). In addition to characterizing the HRQOL of children with SCD-SS, it is important to understand how SCD-SS may change HRQOL over time and impact child development. Much of the SCD literature focuses on health care utilization and pain management (Jacob & Mueller, 2008; Shankar et al., 2008), and does not look at change in HRQOL over time. Therefore, the aims of this study were twofold: (1) to characterize the HRQOL of children with SCD-SS compared to normative

data derived from non-Caucasian US children (Landgraf et al., 1999); and (2) to investigate changes in HRQOL over a 12-month period of participation in a nutritional supplementation study.

Methods

Participants

This study was part of a larger longitudinal randomized controlled trial to determine the effects of daily vitamin A or vitamin A plus zinc supplementation at the Recommended Dietary Allowance (RDA) for age and gender vs. a placebo supplementation on hospitalization frequency (including pain and fever events), nutritional, growth and hematological status over the course of 12 months in children with SCD-SS 2.0 to 12.9 years old. Results for this larger vitamin A supplementation trial are available in detail elsewhere (Dougherty et al., 2012). HRQOL data was collected for children ages 5 years and older at the baseline and 12 month visits as part of this study. There were a total of 62 participants in the study, and 47 had information regarding HRQOL at the baseline protocol visit. All participants in the larger study were diagnosed with SCD-SS and had a suboptimal serum vitamin A status (retinol < 34.5 µg/dL) at screening to be eligible for enrollment. The serum vitamin A criterion was established in support of the larger study of vitamin A status and illness/hospitalization rates for participants. The HRQOL assessment was not obtained for the youngest children (< 5 yrs). Children and parents had 5 protocol visits (baseline, 3, 6, 9, and 12 months), and subjects did not receive behavioral interventions or educational sessions as part of this study. To ensure timely enrollment, we recruited participants at one of five centers, four were affiliated with The Children's Hospital of Philadelphia (CHOP), and the fifth was Newark Beth Israel Hospital in New Jersey (non-CHOP), a collaborating center with similar standard of care. Participants were recruited through their hematologists and visits completed while in their usual state of good health. Children who were prescribed hydroxyurea or transfusion therapy were excluded. The study was approved by the Committee for the Protection of Human Subjects at The Children's Hospital of Philadelphia and the Institutional Review Board at Newark Beth Israel Medical Center. Informed consent was obtained from all participants' parents or guardians, and assent was obtained as appropriate from children aged 7 years old and older.

Hospital Visits

Information on the number of hospital visits for the year during and prior to the study was collected, including day hospital visits (stays less than 24 hours) to the Hematology Acute Care Unit (HACU) and inpatient admissions. These data were identified through hospital medical records, hematology outpatient charts and parental reports.

Child Health Questionnaire Parent Report Form 50 (CHQ)

HRQOL was assessed using the 50-item CHQ-parent-report form for all participants aged 5 and older in order to measure children's physical and psychosocial domains of health. The CHQ is a commonly used generic measure of health related quality of life normed for children aged 5 to 18 years of age (Landgraf et al., 1999). Parent-reported rather than child-reported CHQ was used because the child self-report version is not suitable for children less

than 10 years of age. We chose not to use the child-report for the school age portion of the sample because parent-reported HRQOL may be more reliable than child-reported HRQOL particularly if children are relatively younger (Erhart, Ellert, Kurth, & Ravens-Sieberger, 2009) and sicker, and because the parent form can be completed in less time than the lengthier 87-item child self-report. The CHQ is a valid tool to assess HRQOL in children with SCD (J. A. Panepinto, O'Mahar, DeBaun, Rennie, & Scott, 2004) and has shown good discriminative validity when comparing physical and psychosocial summary scores of children with other chronic illnesses to those scores in normative samples (McCullough & Parkes, 2008). Items cover 14 specific health domains assessing the child's health status over the last four week period. The 14 domains are physical function, role/social limitations due to physical problems, general health perceptions, bodily pain/discomfort, family activities, role/social limitations due to behavioral problems, role/social limitations due to emotional problems, impact on parent time, impact on parent emotions, self-esteem, mental health, general behavior, family cohesion, and change in health. Answers to the questions are reported as levels of intensity or agreement rated on a 4 to 6 point Likert scale. Each scale score is calculated by summing the scale items to receive a total score between 0 and 100, and higher scores indicate better health status. In addition to each of the 14 domain scores, the individual scores can be aggregated to derive two summary scores, one for physical health and one for psychosocial health. The summary scores are transformed into standardized scores with a mean of 50. The CHQ was administered to parents or guardians by trained research staff and data were collected twice, once at the baseline visit and again at the 12-month protocol visit.

Caregivers also reported the child's medical and behavioral co-morbidities as part of the CHQ, and provided details regarding their relationship to the participant, marital status, highest level of education achieved, and job status. Of the 62 participants enrolled in the larger prospective study, parents or guardians of 47 participants completed the CHQ-PF 50 at baseline, and 39 at the 12-month follow-up visit. For all but five participants, the same parent or guardian completed the assessment at both visits.

Growth and Pubertal Status

Anthropometric measurements were obtained in triplicate according to standardized techniques (Lohman, Roche, & Martorell, 1988) and the mean used for analysis. Body mass index (BMI) was calculated (kg/m^2) from weight using a digital scale (Scaletronix, White Plains, NY) and standing height using a stadiometer (Holtain, Crymych, UK) at both baseline and 12 months. Age- and sex-specific height (HAZ), weight (WAZ) and BMI (BMIZ) Z scores were calculated using the Centers for Disease Control and Prevention 2000 reference standards ("Kuczmarski RJ, Ogden CL, Grummer-Strawn LM, Flegal KM, Guo SS, Wei R, Mei Z, Curtain LR, Roche AF, Johnson CL. CDC Growth Charts: United States. Advance data from vital and health statistics; no 314 ed. Hyattsville, MD: National Center for Health Statistics 2000: 1–28.,"). For children eight years of age or older, pubertal status according to the criteria of Tanner (Tanner, 1962) was determined using a validated self-assessment questionnaire (Morris & Udry, 1980), and children in Tanner stage 1 were considered to be prepubertal.

Biochemistry

To determine hematological status, a complete blood count with differential, hemoglobin F (HgbF) and reticulocyte count were assessed using standard techniques at the Clinical Hematology Laboratory and CHOP. Serum retinol was determined by high-performance liquid chromatography (Green Laboratory, Pennsylvania State University, University Park, PA).

Statistical Analyses

Descriptive statistics for total sample for demographic, growth, pubertal, hematological, and retinol status and hospitalizations are presented as mean $\pm$ SD for continuous variables and proportions for categorical variables. Subjects were further classified as “ever hospitalized” or “not hospitalized” in the year prior to the baseline visit. Descriptive statistics for each of the CHQ global scales, summary scales and subscale scores were computed (mean $\pm$ SD). Co-morbidity data from the CHQ were further grouped into broad categories and participants were classified as whether or not they had “any medical co-morbidity” or “any behavioral co-morbidity” in addition to SCD-SS. Caregiver educational achievement was categorized as some high school or diploma vs. any vocational, college or professional education beyond high school.

Although the CHOP affiliated sites and one non-CHOP affiliated site were chosen to have similar standard of care for children with SCD-SS, there may have been differences in the types of care offered at each site. Therefore, differences at baseline and at 12 months in CHQ scale scores between CHOP and non-CHOP participants were examined using independent t-tests. Participants' CHQ scores for each subscale were then compared with normative CHQ data using independent t-tests, a two-sample means comparison test, using the means and standard deviations generated from each sample for each particular subscale. This was performed to assess how the CHQ scale scores in children with SCD-SS at baseline compared with those for a normative sample (n=66) of US African American healthy children (Morris & Udry, 1980). This comparison with the normative sample was then repeated for the CHQ scores obtained for the SCD-SS after participation in the 12-month nutritional supplementation study. Changes from baseline to 12 months in each CHQ subscale were assessed with Wilcoxon signed-rank test for the subjects with both baseline and 12 month follow-up data (n=39).

In children with SCD-SS, differences in CHQ scale scores in the three different supplementation groups (vitamin A, vitamin A plus zinc, and placebo) at baseline and score change over time were examined using analysis of variance (ANOVA). Multiple regression and ANOVA were used to test for significant predictors of the CHQ subscale scores in subjects with SCD-SS at baseline. Possible predictors tested were age, gender, growth and hematological status variables, whether hospitalized in the year prior to the study, presence of medical or behavioral co-morbidities, and caregiver education beyond high school and work status. Power analysis indicated that a minimum sample size of 34 was required to detect a difference of five points (which is considered a clinically meaningful difference (Landgraf et al., 1999) between baseline and 12-month on the CHQ assuming 80% power,

standard deviation of the difference of 10 and alpha of 0.05. All analyses were performed using STATA 9.0 (College Station, TX), and alpha was set at 0.05.

Results

Demographic and growth and health characteristics for the children with SCD-SS are presented in Table 1. The sample was predominantly African American (96%) and pre-pubertal with suboptimal growth and nutritional status, serum retinol levels and a hematological profile typical of children with SCD-SS. All subjects had suboptimal serum retinol status ($<30 \mu\text{g/dL}$) at baseline, and, of these, 64% had retinol in the deficient range ($<20 \mu\text{g/dL}$). In the year prior to the study, two-thirds of the sample had had at least one hospitalization. Caregivers reported medical co-morbidities in 62% of the sample, with asthma predominating, and behavioral co-morbidities in 36% of the sample with attentional and learning problems predominating. Table 2 presents the characteristics of the caregivers who completed the CHQ, reporting for the children. The vast majority of the caregivers were biological mothers. Most were married, had education beyond high school and worked either part or full-time.

We had both CHOP ($n=37$) and non-CHOP ($n=10$) participants with SCD-SS as part of our recruitment pool, and the CHOP and non-CHOP children were similar in all CHQ scales with one exception, the global general behavioral subscale at baseline only (76.9 vs. 91.5, $p=0.04$). Therefore, CHOP and non-CHOP children with SCD-SS were combined. There were no significant differences in baseline scores or change over time based on the larger study experimental condition, i.e. vitamin A supplementation vs. vitamin A and zinc vs. placebo supplementation participants. The comparison of CHQ global scale, summary scale and subscale scores between subjects with SCD-SS at both the baseline ($n=47$) and 12-month visit ($n=39$) and the normative sample of African American healthy children ($n=66$) are presented in Table 3. At baseline, children with SCD-SS had significantly lower scores than the healthy children in the normative sample in the domains of general health, overall physical health (physical health summary score), and parental emotional impact, but a significantly higher score in the domain for mental health. After 12 months of participation in the nutritional supplementation study, CHQ scores for overall physical health and parental emotional impact were statistically the same as the normative scores. The somewhat higher scores for self-esteem and emotional behavioral role function at 12 months in children with SCD-SS compared to the normative sample did not reach statistical significance.

Based on multiple regression analyses, significant predictors of CHQ subscale scores were identified at baseline in the subjects with SCD-SS. Scores for overall physical health were significantly lower in children who had been hospitalized at least once in the year prior to the study compared to those who had not (β -coefficient $\pm$ SE, -7.1 ± 3.1 , $p=0.028$). Scores for overall psychosocial health were significantly lower in children with behavioral co-morbidities compared to those without (-8.4 ± 2.2 , $p=0.0003$). Having a behavioral co-morbidity was also associated with lower scores for parental emotional impact (-25.1 ± 7.2 , $p=0.001$), emotional behavioral role functioning (-15.0 ± 5.2 , $p=0.006$), and impact on parental time (-14.7 ± 5.8 , $p=0.016$). Lower scores for the mental health scale were predicted by both hospitalization (-8.5 ± 3.1 , $p=0.009$) and behavioral co-morbidity (-8.4 ± 0.01),

and lower scores for general health scale were predicted by behavioral co-morbidity (-11.3 ± 5.3 , $p=0.039$) and female gender (-10.8 ± 5.2 , $p=0.044$). Lastly, poorer scores for body pain were more common in females (-12.6 ± 6.3) and in children with lower BMI Z scores (-6.5 ± 3.2 each unit lower Z score, $p=0.04$). Females had lower scores for physical role functioning than males (-14.0 ± 6.3 , $p=0.03$).

Table 4 presents the change in CHQ scores over time in the subjects with SCD-SS. There was a significant improvement in CHQ scores for physical role functioning ($p=0.03$). The improvement in scores for overall physical health was not significant ($p=0.07$). Females had a greater increase in physical role functioning from baseline to 12 months than males ($+11.7 \pm 5.0$, $p=0.02$) with a somewhat greater increase in children with caregivers with educational achievement beyond high school ($+9.6 \pm 4.9$, $p=0.059$).

Discussion

The results of this study indicate that children with SCD-SS in our sample had lower levels of HRQOL than healthy African American children at baseline in three domains: child general health, overall physical health, and parental emotional stress. On the other hand, baseline mental health was higher in children in our sample compared with the reference group. Our results were different from a number of other studies that reported a more consistent trend in poorer HRQOL across a greater number of scale scores for youth with SCD than healthy children, particularly in physical health and pain scale scores (Dale, Cochran, Roy, Jernigan, & Buchanan, 2011; Palermo et al., 2002; J. A. Panepinto et al., 2005; J. A. Panepinto et al., 2004). Unlike the physical scale scores, there is greater inconsistency in the literature regarding the psychological components of HRQOL in SCD. For example, while some studies have demonstrated that children with SCD have increased behavior problems and more psychopathology (Kell, Kliwer, Erickson, & Ohene-Frempong, 1998; Thompson, Gil, Burbach, Keith, & Kinney, 1993; Thompson et al., 1994), lower self-esteem (Hurtig & White, 1986), and poorer family functioning (Nevergold, 1987), other research indicates that most children and families learn to cope with the challenges associated with SCD (Barbarin & Christian, 1999; Lemanek, Moore, Gresham, Williamson, & Kelley, 1986; Midence, McManus, Fuggle, & Davies, 1996). The latter finding is supported by our results that indicate that children with SCD have higher baseline mental health than healthy children. This finding of good/normal mental health is also consistent with reports in children with other medical conditions besides SCD such as end-stage renal disease (Eijssermans, Creemers, Helders, & Schröder, 2004), juvenile idiopathic arthritis (Landgraf et al., 1999; Oliveira et al., 2007) and epilepsy (Landgraf et al., 1999). It has been suggested that individuals with chronic diseases like SCD maintain good mental health as a result of greater social support and a change in how they perceive their condition (Barbarin & Christian, 1999; Schlenk et al., 1998; Sprangers & Schwartz, 1999). Children in our study with SCD-SS may use these factors to bolster their mental health beyond reference values as a protective mechanism in order to cope with their disease. Further, while individuals with SCD may initially have anxiety about their condition, most eventually return to their baseline level of mood (Heady & Wearing, 1992).

Consistent with other pediatric SCD research (Dale et al., 2011; Dampier et al., 2010; Okpala et al., 2002), being hospitalized was significantly related to lower HRQOL compared with no history of hospitalization. Likewise, our result that parent or guardian reported behavioral comorbidity was associated with worse psychosocial HRQOL in children with SCD is also consistent with the literature (J.A. Panepinto, Hoffmann, & Pajewski, 2009; J. A. Panepinto et al., 2005). These results are not unexpected since being hospitalized is disruptive to family life and behavioral problems create family tension. Additionally, we found gender differences indicating that girls had more limited baseline role physical functioning than boys, and that girls in this study significantly increased their role physical functioning more than boys following the intervention. We have reported that children with SCD-SS in this study were shown to have attenuated maximal muscle strength (handgrip strength) and peak power (assessed by vertical jumps using a force plate) at baseline compared to healthy African American children of similar age, gender and pubertal status (Dougherty, Schall, Rovner, Stallings, & Zemel, 2011). This poorer physical conditioning was evident even after controlling for growth and body composition deficits, and may help to account for their poorer CHQ scores in general physical health. Boys and girls had similar actual deficits in muscle strength and power (Dougherty et al., 2011). Palermo and colleagues (Palermo et al., 2002) have also found lower physical functioning in girls than boys with SCD, and have suggested that parents may perceive that girls may not be able to participate in physical activities at the same level as boys. Interestingly, and consistent with our data, Palermo and colleagues (Palermo et al., 2002) indicate that girls did not have higher rates of hospitalizations, or comorbidities that could account for this gender difference. In our sample, girls did not have poorer physical conditioning than boys and both boys and girls had suboptimal growth and nutritional status and suboptimal vitamin A status which did not improve with vitamin A supplementation (Dougherty et al., 2012). It is unclear why females reported greater bodily pain scores than males in our study. There may be undocumented comorbidities associated with female's poorer general health scale score that may be associated with higher pain in females than males. Another finding of our study was that higher caregiver education was significantly associated with greater change in the role physical functioning scale score following the intervention. A similar finding was reported by others who have found higher parental education to be associated with better psychosocial HRQOL (Palermo, Riley, & Mitchell, 2008; J. A. Panepinto et al., 2005).

This is the first known study to compare HRQOL in SCD-SS with norms for non-Caucasian U.S. healthy children. We examined longitudinal changes in HRQOL following participation in a 12 month nutritional intervention. Interestingly, the role functioning physical scale was the only scale to significantly increase over the intervention. Other scale scores did not change over time. Future research should examine potential mediators of change in HRQOL (e.g., coping, family functioning, disease-related stress) in order to advance understanding of HRQOL variables in this population. Information for potential mediators was not collected as part of our study, and the role of these mediators may partly explain why a difference was not identified in scale scores other than the role functioning physical scale. It is unclear if the increase in the role functioning physical scale was the direct result of being study volunteers, the specific intervention, or due to changes in normal development. Since this study did not collect specific information to potentially explain the

mechanisms behind changes in HRQOL, the exact reasons for improvement cannot be determined. Likewise, the nature of this study design limits any conclusions about the cause and effect relationship between variables. Future research should explore potential mediators of change in HRQOL (e.g., coping, family functioning, disease-related stress) in order to advance understanding of HRQOL variables in this population. There were no improvements in growth and nutritional status, including vitamin A status in these children after 12 months of supplementation (Dougherty et al, 2012), and there were no differences in HRQOL trends among the three randomization groups, suggesting that taking vitamin A supplements per se was not responsible for these results. Other than a study examining changes in HRQOL in children with SCD with an acute vaso-occlusive painful event cared for in an emergency room setting (Brandow, Brousseau, Pajewski, & Panepinto, 2010), there are no other reports in SCD of the responsiveness of HRQOL measures over time. Further research is needed in order to determine interventions for improving HRQOL in children with SCD and whether a valid and responsive disease specific measure of HRQOL for children with SCD is needed.

There are some limitations to consider when examining the results of this study. First, we excluded subjects with SCD-SS who were on hydroxyurea medication or chronic blood transfusion therapy. These therapies are becoming more common in children and adolescents with SCD. Thus, our study sample may have been healthier compared to children with SCD-SS now treated with either hydroxyurea or chronic transfusions. This may explain in part why many of the scale scores of HRQOL for our children were not significantly different from the healthy African American comparison group. Further the demographic characteristics for the African American comparison group may be different than the characteristics of the children we studied. This potential difference could also explain why differences in HRQOL were not larger. Also, caregiver proxy report was used for assessing HRQOL. Previous research indicates that parent and child reported HRQOL differ, with parents of children who are chronically ill generally reporting poorer HRQOL than children (J. A. Panepinto et al., 2005; Sawyer, Antoniou, Toogood, & Rice, 1999; Wake, Hesketh, & Cameron, 2000). It may be that caregiver distress over the child's SCD may result in overreporting of severity of their disease in HRQOL. Thus, if we had assessed children's self-reported HRQOL, our results may have been different. Further, it is possible that parent reports of children's HROL could have changed over time regardless of the intervention. Including both child and parent reports of HRQOL and exploring potential reasons for discrepancies between each report as well as accounting for potential parental reporting change over time are areas for future research. Additionally, since the CHQ is designed for children and adolescents in the age span of 5–18, age-related differences in scores are likely to occur. However, we attempted to account for this in part by including age as a covariate in regression models when testing for significant predictors of the CHQ subscale scores. Another limitation of our study is that a generic, broadly applicable measure rather than disease-specific measure was used to assess HRQOL. There has been only one known validation study of the CHQ for use in children with SCD that examined the relationship between severity of disease and HRQOL (J. A. Panepinto et al., 2004). There is a need for further validation of this tool to determine whether it captures specific aspects unique to the HRQOL of children with SCD, and that may help differentiate HRQOL not

only by SCD severity but also from other chronic health conditions. This may help determine whether a specific tool for measuring HRQOL in children with SCD is needed, as exists for children with other chronic conditions that include epilepsy (Sabaz et al., 2000), obesity (Modi & Zeller, 2008), and cystic fibrosis (Quittner, Buu, Messer, Modi, & Watrous, 2005).

It is important that care providers such as primary care nurse practitioners, psychologists, health educators, social workers, etc. have an understanding of HRQOL of children with SCD in order to best manage the health problems they experience. Through health education and health promotion strategies aimed at helping patients take control of their condition and at identify social supports, these care providers play a key role in helping the child and family better manage their SCD. Our results suggest that children with SCD have more limitations in overall general health while maintaining normal levels of mental health. Good mental health, coupled with strong family cohesion, productive family activities, and mature child behavior may have helped children with SCD in our study better maintain higher levels of physical function and cope with their disease compared with children in other studies that have poorer psychosocial health. Testing whether providing social support and behavioral coping skills helps improve overall HRQOL in children with SCD could be an area for future research.

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Table 1**Characteristics of Subjects with SCD-SS at Baseline**

	(<i>n</i> = 47)
Age, years	8.6 ± 2.4
Gender, female	20 (43%)
Ethnicity, Hispanic	3 (6%)
Race	
African American	45 (96%)
Caucasian	2 (4%)
<i>Growth and Pubertal Status</i>	
Height-for-age Z score	−0.6 ± 0.8
Weight-for-age Z score	−0.8 ± 1.0
BMI-for-age Z score	−0.6 ± 1.0
Pre-pubertal (Tanner stage 1)	41 (87%)
<i>Hematological and retinol status</i>	
Hemoglobin, g/mL	8.1 ± 0.9
Hematocrit, %	23.5 ± 2.9
HgbF, %	8.3 ± 5.3
Reticulocytes, %	13.6 ± 4.6
Serum retinol, ug/dL	18.1 ± 4.0
<i>Hospitalizations, year prior to study</i>	
Total hospitalizations	1.7 ± 1.8
Ever hospitalized	31 (66%)
<i>Medical Co-Morbidities</i>	
Any	29 (62%)
Asthma	14 (30%)
Chronic Allergies	8 (17%)
Orthopedic/Bone/Joint problems	3 (6%)
Chronic respiratory problem (non-asthma)	5 (11%)
Sleep disorder	5 (11%)
Deafness	1 (2%)
Vision	7 (15%)
Other	5 (11%)
<i>Behavioral Co-morbidities</i>	
Any	17 (36%)
Attentional problems	10 (21%)
Behavioral problems	4 (9%)
Developmental delay or mental retardation	3 (6%)
Learning problems	11 (23%)
Speech problems	3(6%)

Values are mean ± SD for continuous variables, or n(%) for categorical variables Hospitalizations include HACU visits (Hematology Acute Care Unit, <24 hours) and hospitalizations lasting > 24 hour visits).

Table 2

Characteristics of Caregiver Completing the CHQ

	<i>(n = 47)</i>
<i>Gender, female</i>	44 (94%)
<i>Relationship of caregiver to child</i>	
Biological parent	44 (94%)
Guardian	1 (2%)
Adoptive parent	1 (2%)
Other	1 (2%)
<i>Caregiver marital status</i>	
Married	26 (55%)
Divorced/separated	2 (4%)
Never married	18 (38%)
Not reported	1 (2%)
<i>Caregiver education</i>	
Did not graduate from high school	6 (13%)
High school diploma	16 (34%)
Vocational school/some college	12 (26%)
College degree	12 (26%)
Professional/graduate degree	1 (2%)
<i>Caregiver job status</i>	
Working part- or full-time	34 (72%)
Not working	13 (28%)

Table 3

Child Health Questionnaire scale scores for sample versus Non-Caucasian normative data

CHQ Scales	Baseline Score mean, (SD) n = 47	12 Month Score mean, (SD) n = 39	Normative sample ^a mean, (SD) n = 66	Baseline vs. Norm p value	12 Month vs. Norm p value
Global General Health	72.7 (21.1)	70.1 (22.3)	-		
Global General Behavior	80.0 (20.1)	75.4 (20.6)	-		
General Health	50.0 (18.8)	51.1 (15.6)	68.0 (18.9)	<.001	<.001
Bodily Pain	74.0 (22.6)	75.6 (20.4)	81.4 (20.8)	.075	.167
Self-esteem	85.3 (17.0)	86.5 (18.0)	80.7 (12.3)	.098	.053
Physical Functioning	87.4 (23.2)	92.5 (19.3)	91.7 (21.4)	.312	.848
Physical Health Summary	44.7 (10.6)	47.6 (9.5)	50.2 (12.3)	.015	.259
Psychosocial Health Summary	52.3 (8.1)	53.0 (7.5)	50.1 (9.8)	.210	.115
Role functioning – Physical	89.7 (22.1)	95.3 (14.3)	87.9 (26.4)	.703	.109
General Behavior	73.4 (15.4)	72.9 (15.8)	73.7 (17.9)	.926	.818
Role Emotional - Behavioral	91.3 (18.4)	95.4 (14.6)	87.0 (25.4)	.324	.062
Family Activities	82.5 (19.5)	84.2 (20.8)	82.8 (25.4)	.946	.751
Mental health	82.9 (10.7)	82.4 (13.5)	76.7 (15.5)	.020	.059
Family Cohesion	72.7 (25.8)	73.2 (18.0)	71.1 (21.9)-	.723	.614
Parental Impact - Emotional	67.0 (26.5)	73.1 (23.9)	77.9 (23.0)	.022	.311
Parental Impact - Time	83.2 (20.3)	86.6 (25.9)	79.9 (27.8)	.490	.224

^aLandgraf et al., (1999)

Table 4

Baseline and 12-Month CHQ Scale scores (n = 39)

CHQ Scales	Baseline Score	12-Month Score	p-value
Global General Health	74.6 (22.3)	70.1 (22.3)	0.424
Global General Behavior	79.5 (21.1)	75.4 (20.6)	0.239
General Health	51.8 (17.9)	51.1 (15.6)	0.660
Bodily Pain	76.4 (22.5)	75.6 (20.4)	0.794
Self-esteem	86.3 (18.0)	86.5 (18.0)	0.675
Physical Functioning	86.8 (24.8)	92.5 (19.3)	0.263
Physical Health Summary	45.1 (10.8)	47.6 (9.5)	0.070
Psychosocial Health Summary	52.7 (8.3)	53.0 (7.5)	0.748
Role functioning – Physical	89.3 (22.1)	95.3 (14.3)	0.033
General Behavior	73.3 (14.9)	72.9 (15.8)	0.978
Role Emotional - Behavioral	91.2 (19.1)	95.4 (14.6)	0.310
Family Activities	82.1 (19.9)	84.2 (15.8)	0.667
Mental health	83.5 (11.0)	82.4 (14.6)	0.877
Family Cohesion	71.9 (27.2)	73.2 (20.8)	0.697
Parental Impact - Emotional	66.9 (28.3)	73.1 (13.5)	0.198
Parental Impact - Time	85.5 (18.8)	86.6 (25.9)	0.507

Significance determined using Wilcoxon signed rank test to adjust for skewness