Learning outcome, instructions for completing the evaluation, and statements of disclosure can be found on page 118.

Depression, Anxiety, and Quality of Life In Children and Adolescents With Sickle Cell Disease

J. Kelly Graves, Christopher Hodge, and Eufemia Jacob

The relationships among depression, anxiety, and quality of life were tested, as were the effects of age, gender, and pain frequency on these variables in children (n = 44) and adolescents (n = 31) with sickle cell disease. Participants completed the Revised Child Anxiety and Depression Scale (RCADS) and the Pediatric Quality of Life (PedQL Generic Model). The mean and standard deviation for summary RCADS scores for the majority of participants were below the clinical thresholds of $T < 65$, indicating low risk for depression (n = 65; 89.3%) and anxiety (n = 70; 93.3%). The subscale scores for the different dimensions of QOL health were a) psychosocial (73.3 ± 15.9), b) emotional (75.0 ± 20.7), c) social (80.8 ±19.1), d) school functioning (64.0 ≥ 19.8), and e) physical (77.4 ± 17.4). Significant negative correlations were found between mean total quality of life scores and symptoms of a) general anxiety (r = -0.51, $p < 0.0001$), b) depression (r = -0.66, $p < 0.0001$), c) obsessive compulsive (r = -0.53, $p < 0.0001$), d) panic (r = -0.60, $p < 0.0001$), and e) social phobia (r = -0.57, $p < 0.0001$). Age and gender did not have significant effects on risk for depression and anxiety or poor QOL. Pain frequency also did not have significant effects on the risk for depression and anxiety. Findings suggest that health care providers need to screen for anxiety and depression, and make referrals for early interventions to improve quality of life and promote school function in youth with sickle cell disease.

Children and adolescents with sickle cell disease may be at risk for psychological distress, especially depression and anxiety, as they face the challenge of reoccurring painful exacerbations or crises, which can be life-threatening (Benton, Boyd, Ifeagwu, Feldtmose, & Smith-Whitley, 2011; Carpentier, Elkin, & Starnes, 2009; Simon, Barakat, Patterson, & Dampier, 2009). Sickle cell disease, a genetic disease, is characterized by painful vaso-occlusive events when sickle-shaped erythrocytes block blood flow to cells, organs, and bones (Bryant, 2011). Of all the negative effects and harm of unrelieved pain, the greatest may be a poor quality of life (Benjamin, 2008), in part from emergency care and hospitalizations, which interrupt family life, school attendance, and social activities. It was hypothesized that the frequent episodic exacerbations of pain may increase the incidence of depression and anxiety, ultimately affecting the health-related quality of life in children and adolescents with sickle cell disease.

The research questions were 1) to what extent are children and adolescents with sickle cell disease at risk for depression and anxiety, 2) what is the quality of life in children and adolescents with sickle cell disease, and 3) what factors (age, gender, and number of pain episodes) have significant effects on risk for depression and/or anxiety and quality of life in children and adolescents with sickle cell disease?

Review of the Literature

The Conceptual Model of Health-Related Quality of Life (Ferrans, Zerwic, Wilbur, & Larson, 2005; Wilson & Cleary, 1995) provided the context for this research. In this model, health-related quality of life (HRQoL) was proposed to have a reciprocal relationship to an individual's perceptions of health, functional status, symptoms, and biological function (Ferrans et al., 2005). In addition, characteristics of the individual and environment are linked to these variables.

The physical environment affecting HRQoL may include neighborhoods, home, and schools (Palermo, Riley, & Mitchell, 2008). For example, neighborhood poverty levels indicating socioeconomic distress, unemployment, female head of the household, and number of high school dropouts were factors that could affect HRQoL. Poverty at the individual and neighborhood/community levels was a predictor of children's functional disability and negative physical and psychosocial HRQoL. Panepinto, Pajewski, Foerster, Sabnis, and Hoffmann (2009) also found that when comparing the impact of family income of 104 children with severe sickle cell disease as compared with 74 children without the disease, those with sickle cell disease who had a lower family income had a significantly worse overall HRQoL.

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The social environment affecting HRQoL may include the influence of family, friends, and health care personnel. In a study of perceptions of 1,772 children with sickle cell disease compared with a parent/caregiver, both children and parent/caregiver reported that school functioning decreased in response to asthma and pain (Dampier et al., 2010). Children also cited the negative effect of sickle pain and asthma on sleep/fatigue scales, while parents/caregivers reported sickle pain and avascular necrosis (cellular death of bone) had a negative effect on sleep/fatigue scales. The similarity of responses suggests that perceptions of serious events are shared, and a parent/care giver may have influenced the child.

In addition to physical and social environmental characteristics, the unique characteristics of the individual, such as age, sex, and ethnicity, developmental stage in the life span, and psychological and biological factors, may affect HRQoL. Biological factors, such as skin color, sex, ethnicity, and an inherited risk of sickle cell disease, are considered unchangeable. However, psychological factors are considered modifiable if health care managers are aware of the impact of chronic pain on a child. A relationship was found between children with sickle cell disease and the frequency and number of psychological disorders, especially depression and anxiety (Barakat, Schwartz, Simon, & Radcliffe, 2007; Myrvik, Campbell, Davis, & Butcher, 2012).

In yet another retrospective study of 2,194 children and adolescents with sickle cell disease, 46% were diagnosed with a depressive disorder (Jerrell, Tripathi, & McIntyre, 2011). Ninety percent of those with depression had dysthymia diagnosed at age 9 years, and the other 10% had a major depressive disorder diagnosed at age 14 years (Jerrell et al., 2011). This cohort with depression experienced more pain and organ damage complications, and incurred more health care costs as a result of emergency care. Further, Benton and colleagues (2011) found that despite improved medical advances that address anemia, infection, and sickle cell crises, adolescents are still at risk for depression and anxiety. These authors recognized that awareness of these risks assist health practitioners in identifying depression and anxiety, and implementing early interventions (Benton et al., 2011).

Unal, Toros, Kutuk, and Uyaniker (2011) found a relationship between the number of pain episodes and depression. Depression levels of 40 children with over 10 pain episodes were higher than those who had 1 to 4 episodes in a year. These studies are consistent with findings by others (Barakat et al, 2007; Gil et al., 2003), who found an association between adolescents who reported higher pain, frequency, and intensity with increased symptoms of depression and anxiety. It is clear that anticipating pain, possible hospitalization, and an interruption of daily life has an effect on the children and adolescents with sickle cell disease.

In addition to medical and neuropsychological problems, children and adolescents with sickle cell disease are at risk for social and academic/vocational issues included in the quality of life (Lemanek & Ranalli, 2009). Previous studies also have reported higher school absence among children with sickle cell disease and reported poor academic achievement (Knight-Madden, Lewis, Tyson, Reid, & Moo Sang, 2011; Logan, Simons, Stein, & Chastain, 2008). A chronic, debilitating illness may interfere with peer group identification, especially at school, when conforming to the group norm is a developmental stage for a pre-adolescent or adolescent student (Santrock, 2015). Attempts to identify with peers may be further challenged when physical and sexual maturation is delayed. Although an adolescent can expect to achieve normal height, weight, and sexual function as an adult, these delays may become embarrassing for a teenager (Heeney & Dover, 2009; Redding-Lattinger & Knoll, 2006). Additionally, chronic complications of sickle cell disease become more pronounced in adolescence, such as renal and cardiopulmonary dysfunction and avascular necrosis of the shoulders and hips (Castro & Gladwin, 2005; Dampier et al., 2010). These complications further set a student with sickle cell disease apart from others, and a reduced quality of life may result in time lost from academic, vocational, and social life.

Hoff, Palermo, Schluchter, Zebracki, and Drotar (2006) found that girls reported more pain intensity than boys in a one-year sickle cell disease study. Older girls (10 years and older) also reported more pain, medication use, and restrictions than boys in daily life (Ross-Isigkeit, Thyen, Stoven, Swartzenberger & Schmucker, 2005). Girls also scored significantly lower in general quality of life and function scales (Dampier et al., 2010). Increased age in adolescents between 12 and 19 years was significantly related to anxiety and depression (Carpentier et al., 2009). Studies suggest that pain and complications from sickle cell disease increase from birth to adulthood, and that depression of children poses a risk factor for later pain and functional disability (Hoff et al., 2006; Perquin et al., 2003). Although previous research suggests that effects of sickle cell disease may be gender- or age-specific, research finding reaching significance is limited.

Unpredictable exacerbations of pain and constant vigilance to prevent reoccurrences before adulthood render a child or adolescent vulnerable for depression and anxiety (Barakat et al., 2007; Benton et. al., 2011; Ingerski et al., 2010; Simon et al., 2009). There is evidence that depression in children will occur again in adolescence and in adulthood, with more negative outcomes for those diagnosed as adolescents (Schulenberg & Zarrett, 2006).

This study was designed to address gaps of knowledge regarding selected variables of gender, age, and pain as predictors of depression and anxiety for youth with sickle cell disease. Research is not consistent in suggesting sex and age are predictors of depression and anxiety or quality of life for youth with sickle cell disease. Although studies indicate there may be a relationship between the amount of pain and depression and anxiety, their impact on the quality of life of children and adolescents with sickle cell disease needs further evaluation. Identifying what aspect of the quality of life is perceived as unsatisfactory may be the first step toward a modifiable intervention.

Methods

Design

The research was part of a larger study that examined pain and symptoms in children and adolescents with sickle cell disease. A longitudinal design was used when they were completing a web-based electronic diary related to pain and symptoms during a three-month period (Jacob et al., 2012; Jacob, Duran, Stinson, Lewis, & Zeltzer, 2013; Jacob, Pavlish et al.,

2013). The data presented here were collected as part of baseline assessments when participants completed psychosocial measures that included 1) Revised Child Anxiety and Depression Scale (RCADS) and 2) the Pediatric Quality of Life (PedsQL). The Institutional Review Board at the University of California Los Angeles approved all study procedures.

Sample and Setting

Participants were recruited through the Sickle Cell Disease Foundation of California, a community-based organization that serves approximately 2,000 individuals with sickle cell disease in Southern California. A special program coordinator distributed study flyers and scheduled eligible participants for a two-hour information session, including consenting and enrollment procedures. Participants met the following criteria for inclusion in the study: 1) known diagnosis of sickle cell disease (HgbSS, HgbSC, Hgb Beta Thal); 2) age between 10 and 17 years; 3) ability to read, speak, and write English; 4) ability to use a computer; and 5) parent/legal guardian availability to consent. Participants were excluded if the primary care provider previously informed parents that the child had neurological or cognitive impairments that would hinder completion of outcome measures. Sample size calculation for the larger study was determined by power analyses using GPOWER (Erdfelder, Faul, & Buchner, 1996), indicating that an overall total sample size of 75 participants would yield an effect size of 0.20 in the pain, depression, anxiety, and quality of life measures, with alpha of 0.05 and power of 0.80.

Procedures

Participants and a parent/caregiver completed a demographic questionnaire that included questions about the number of pain episodes experienced at home and/or requiring hospitalizations the previous 12 months. Participants also completed a battery of psychosocial measures using a computer that included measures on anxiety, depression, and quality of life. Data collection procedures occurred in a community site arranged by the Sickle Cell Disease Foundation, and took 45 to 60 minutes to complete.

Instruments

Anxiety and depression. The Revised Child Anxiety and Depression Scale (RCADS) (Chorpita, Moffitt, & Gray, 2005; Chorpita, Yim, Moffitt, Umemoto, & Francis, 2000) is a self-report questionnaire for youth used to measure the risk for depression and anxiety. RCADS is a 47-item self-report form, with a 4-point Likert scale ranging from *never* to *always*. It is designed to assess the rating of each subscale in accordance with the *Diagnostic and Statistical Manual of Mental Disorders*, Fourth Edition, Text Revision (DSM-IV TR) (American Psychiatric Association [APA], 2000). The questionnaire consists of six subscales: Generalized Anxiety Disorder (GAD), Major Depressive Disorder (MDD), Obsessive-Compulsive Disorder (OCD), Panic Disorder (PD), Social Phobia (SP), and Separation Anxiety Disorder (SAD). Raw subscale scores were converted into *T* scores based on age and gender (Chorpita et al., 2005). Cutoff scores were made at $T \geq 65$ to determine whether individuals met DSM-IV TR criteria for risk of anxiety or depressive disorders. Good internal consistency and test-retest reliability have been demonstrated in 109 adolescent males (44.3%) and 137 adolescent females (55.7%); mean age 12.20 years ± 2.89 (range=8.33 ± 18.33 years). Alpha coefficients were SP, $\alpha = 0.81$; PD, $\alpha = 0.85$; GAD, $\alpha = 0.80$; MDD, $\alpha = 0.76$; SAD, $\alpha = 0.78$; and OCD, $\alpha = 0.71$ (Chorpita et al., 2000).

Quality of life. The Pediatric Quality of Life Instrument (PedsQL, Generic Module) was selected to measure quality of life. The scale was developed to assess both adolescent and parent perceptions of health-related quality of life in children with serious health conditions. It measures the core physical, mental, and social health dimensions as well as the school functioning role in children and adolescents. The questionnaire consisted of questions designed to measure the extent of a problem: (0 = never a problem; 1 = almost never a problem; 2 = sometimes a problem; 3 = often a problem; 4 = almost always a problem). Items are reverse-scored and linearly transformed from a 0 to 100 scale (0 = 100, 1 = 75, 2=50, 3 = 25, 4 = 0). The higher PedsQL scores indicate a better health-related quality of life (Varni, Seid, & Kurtin, 2001). The internal consistency reliability for child/self report and parent proxy report ranged from 0.66 to 0.93; the parent/child concordance intercorrelations ranged from 0.305 to 0.944 (Varni et al., 2001). The PedsQL was completed on the day of enrollment and also at the end of the study during a scheduled clinic visit.

Data Analyses

Data were analyzed in a Statistical Package for Social Sciences (SPSS version 20.0, Armonk, NY). Descriptive statistics (frequencies, means, standard deviations) were used to describe demographics, and anxiety and depression (RCADS) and quality of life (PedsQL) scores. Pearson correlations were used to examine the relationships among anxiety, depression, and quality of life scores. Bivariate and multivariate analyses were used to examine factors (age, gender, pain, and sickle cell disease diagnoses) that may have effects on anxiety, depression, and quality of life.

Results

Participants were children and adolescents between 10 and 17 years; mean age 13.0 years, SD 1.9 years; 37 (49.3%) males; and 38 (50.7%) females (see Table 1). The majority of participants had either HgbSS ($n = 35$; 46.7%) or Hgb SC ($n = 23$; 30.7%). Few had Hgb Beta Thalassemia ($n = 7$; 9.3%), and the others were unknown ($n = 10$; 13.3%). The majority ($n = 49$; 73.1%) reported 0 to 2 pain episodes per year that required hospitalization. Participants ($n = 18$; 26.9%) reported more than 3 pain episodes per year requiring hospitalization.

Depression and Anxiety

The mean and standard deviation for summary RCADS scores for the majority of participants were below the clinical thresholds of $T < 65$, indicating low risk for depression ($n = 65$; 89.3%) and anxiety ($n = 70$; 93.3%). However, there were participants with clinical thresholds of $T \geq 65$ (see Table 2). Age (see Figure 1), sex (see Figure 2), and pain frequency did not have significant effects on the risk for anxiety or depression.

Quality of Life

Total mean quality of life scores were 74.7 ± 14.8 on a scale of 0 to 100 (range 34.8 to 100) (see Figure 3). The subscale scores for the different dimensions of quality of life were 1) psychosocial health (73.3 ± 15.9), 2) emotional (75.0 ± 20.7), 3) social (80.8 ± 19.1), 4) school functioning (64.0 ≥ 19.8), and 5) physical (77.4 ± 17.4). Significant negative correla-

Table 1.
Demographics (*N* = 75)

	n (%)
Age	
Children	44 (58.7%)
Mean	11.6 ± 1.1 years
Range	10 to 13 years
Adolescents	31 (41.3%)
Mean	14.9 ± 0.9 years
Range	14 to 17 years
Sex	
Males	37 (49.3%)
Females	38 (50.7%)
Hemoglobin Genotype	
Hgb	35 (46.7%)
Hgb	23 (30.7%)
HgbB	7 (9.3%)
Unknown	10 (13.3%)
Number of Acute Pain Episodes*	
0 to 2 per year	49 (73.1%)
≥ 3 per year	18 (26.9%)

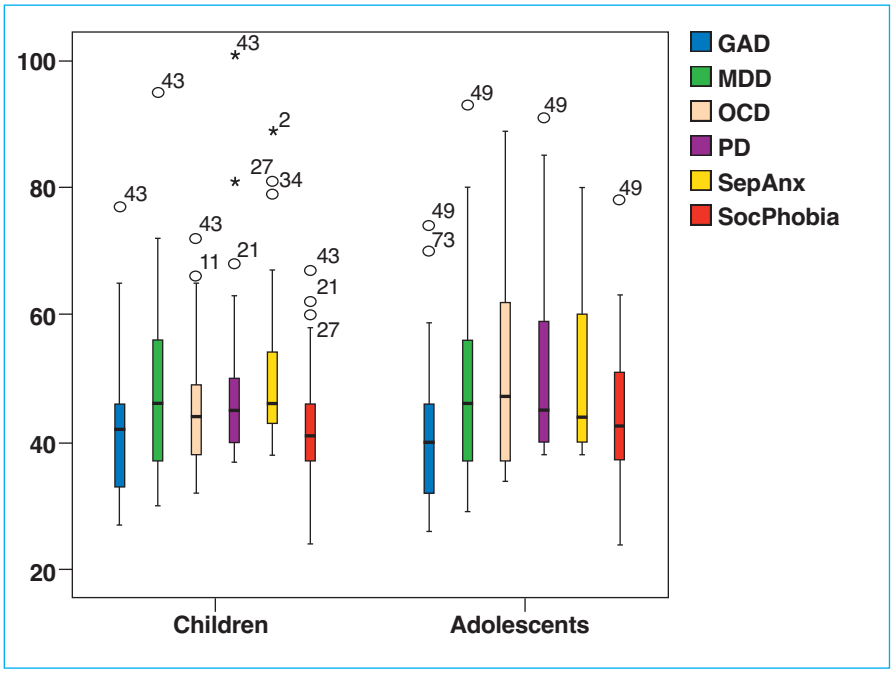
*Number of acute pain episodes requiring hospitalizations.

Table 2.
Revised Child Anxiety and Depression Scale (RCADS) Scores

RCADS	M ± SD* (*T* < 65)	M ± SD** (*T* ≥ 65)
Generalized Anxiety Disorder	42.16 ± 11.10	72.5 ± 5.2
Major Depressive Disorder	47.40 ± 13.71	76.0 ± 12.1
Obsessive Compulsive Disorder	46.67 ± 11.91	73.0 ± 8.8
Panic Disorder	49.47 ± 13.89	79.8 ± 11.6
Social Phobia	42.08 ± 10.47	72.5 ± 7.8
Separation Anxiety Disorder	49.76 ± 11.39	75.7 ± 8.8

*Mean raw scores.
**Cutoff scores were made at *T* ≥ 65 and determine whether individual meet DSM-IV TR criteria for risk of the anxiety or depressive disorders listed.

Figure 1.
Revised Child Anxiety and Depression Scale Scores by Age



Notes: GAD = General Anxiety Disorder, MDD = Major Depressive Disorder, OCD = Obsessive Compulsive Disorder, PD = Panic Disorder, SepAnx = Separation Anxiety Disorder, SocPhobia = Social Phobia.

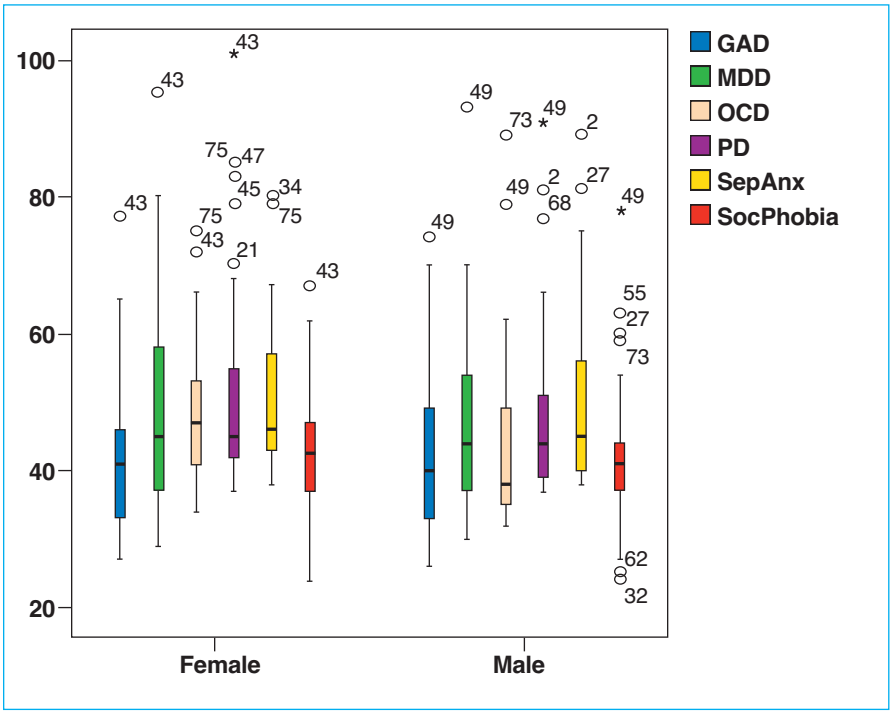
tions were found between mean total quality of life scores and symptoms of 1) general anxiety (r = -0.51, p < 0.0001), 2) depression (r = -0.66, p < 0.0001), 3) obsessive compulsive (r = -0.53, p < 0.0001), 4) panic (r = -0.60, p < 0.0001), and 5) social phobia (r = -0.57, p < 0.0001). Age, sex, and pain frequency did not have significant effects on quality of life scores.

Discussion

It was hypothesized that painful, episodic exacerbations associated with chronic sickle cell disease may increase the vulnerability of children and adolescents for depression and anxiety, and have an impact on their quality of life. Significant negative correlations were found between the total quality of life scores and symptoms of general anxiety, depression, obsessive compulsive disorder, panic, and social phobia. Similar to findings by Palermo, Schwartz, and Drotar (2002), a reduced quality of life was found, particularly for the subscale on school functioning. Functioning pertains to physical, mental, and social activities, especially during school time. Included in the assessment are feelings of sadness, anger, and fear; getting along with others; and keeping up with school work despite absenteeism because of health related issues. Previous studies have found that children with sickle cell disease were able to cope with diverse learning in schools (Ogunfowora, Olanrewaju, & Akenzua, 2005). However, compared to peers, a decrease in social and school competence for children with sickle cell disease was found, but there was no association with disease severity (Barakat, Patterson, Daniel, & Dampier, 2008).

Pain frequency did not have a significant effect on quality of life in children and adolescents with sickle cell disease in our study, which is consistent with previous findings (Barakat et al., 2008). However, others (Schlenz, Schatz, McClellan, & Roberts, 2012) found that the frequency of pain predicted a decrease in the total quality

Figure 2.
Revised Child Anxiety and Depression Scale Scores by Gender



Notes: GAD = General Anxiety Disorder, MDD = Major Depressive Disorder, OCD = Obsessive Compulsive Disorder, PD = Panic Disorder, SepAnx = Separation Anxiety Disorder, SocPhobia = Social Phobia.

Figure 3.
Psychosocial Health, Physical Health, School Functioning, and Quality of Life Scores

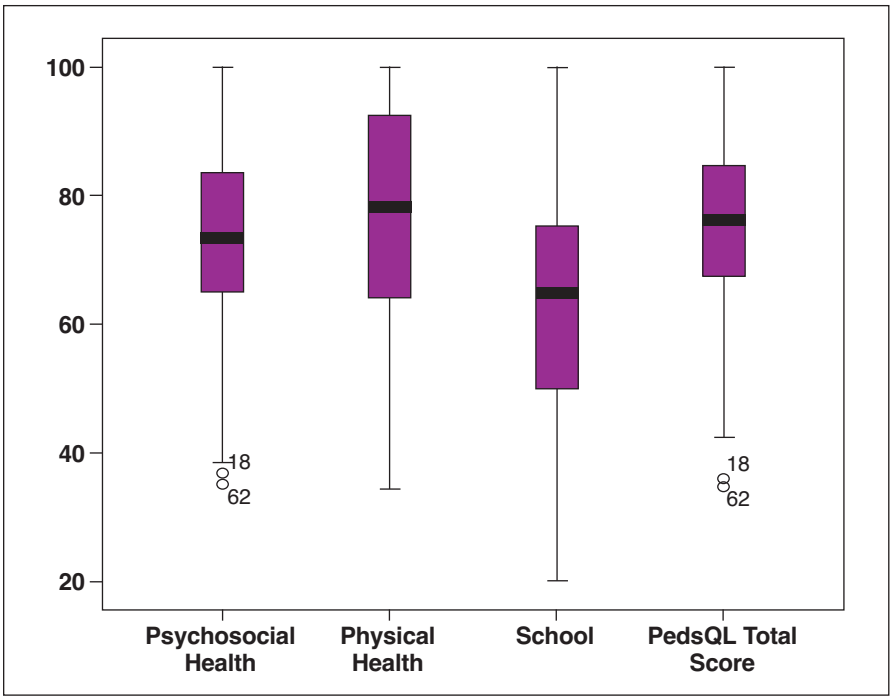


Table 3.
Correlations Between Revised Child Anxiety and Depression Scale (RCADS) Subscales and Quality of Life (QOL) Scores

RCADS	QOL
Generalized Anxiety Disorder	-0.512*
Major Depressive Disorder	-0.662*
Obsessive Compulsive Disorder	-0.534*
Panic Disorder	-0.601*
Social Phobia	-0.569*
Separation Anxiety Disorder	0.146

**Correlation is significant at the 0.0001 level (2 tailed).

of life, which may be attributed to the difference in the methods (self-report versus medical chart reviews) used for assessment of pain frequency.

RCADS scores indicated that the majority of respondents were below the clinical thresholds for depression, although some participants had scores indicating a risk for depression. Scores indicating a low risk for depression may be attributed to a repressive adaptive style where a person with a chronic illness exhibits a defensive coping style, which may camouflage depression and trait anxiety (Phipps & Srivastava, 1997).

The majority of RCADS scores were below the clinical thresholds for anxiety, indicating low risk. However, a few participants had elevated clinical thresholds, indicating a risk for panic disorder, separation anxiety disorder, obsessive compulsive disorder, general anxiety disorder, and social phobia. These prevalence rates for anxiety are higher than the 2% to 4% rates for adolescents in the general population for anxiety disorders (Polanczyk, De Lima, & Horta, 2007). Simon and colleagues (2009) speculated that the depression and anxiety scores that were within normal clinical ranges in their study of children with sickle cell disease could be attributed to the resilience factor. Children may develop psychosocial skills themselves, which serve as an intervention unique to each person rather than other-directed.

The two demographic variables of age and gender did not have signif-

Instructions For Continuing Nursing Education Contact Hours

Depression, Anxiety, and Quality of Life in Children and Adolescents with Sickle Cell Disease

Deadline for Submission: **June 30, 2018** **PED 1603**

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1. To obtain CNE contact hours, you must read the article and complete the evaluation through the ***Pediatric Nursing*** **website** at **www.pediatricnursing.net/ce**
2. Evaluations must be completed **online** by June 30, 2018. Upon completion of the evaluation, your CNE certificate for **1.3** contact hour(s) will be mailed to you.

Learning Outcome

After completing this learning activity, the learner will be able to assess risk for anxiety and depression and the need for early interventions to improve quality of life and promote school function in children with sickle cell disease.

Fees — *Subscriber:* FREE *Regular:* $20

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icant effects on the risk for depression and anxiety. However, pain frequency had statistically significant effects, but the difference was not clinically significant. These findings are consistent with research findings by others who found an association between adolescents who reported higher pain, frequency, and intensity with increased symptoms of depression and anxiety (Barakat et al., 2007; Gil et al., 2003). Simon and colleagues (2009) also found that when the severity of a disease is mild, the incidence is low for depression and anxiety.

Several limitations need to be mentioned when interpreting the study. First, the sample size was small, which limits the generalizability of the findings. Second, convenience sampling in one community in the southwestern United States was used, which limits representativeness to other children with sickle cell disease in other settings. Third, we were not able to collect environmental factors that may contribute to psychological issues. Socioeconomic distress and the lack of social support systems may contribute to depression, anxiety, and quality of life.

Clinical Implications

Nurses and other health care providers do not routinely assess children and adolescents at risk for anxiety and depression. In schools, it might be the teacher who spends the most time with the students and is instrumental in noting changes in psychosocial functioning. Due to the low level of school functioning for participants with sickle cell disease in this study, this could be addressed by enrolling a student in the Individualized Education Program (IEP). The IEP, mandated by the Individuals with Disabilities Act, gives a child who qualifies the opportunity to participate in a traditional classroom situation to meet educational goals through a multi-disciplinary approach. There are several members of the Individual and Education Plan (IEP) team, which include the student, parents or guardian, case manager/teacher, school psychologist, school nurse, and school resource expert. This team approach supports the child with sickle cell disease with important primary prevention interventions. School nurses need to implement early interventions, working with school teachers, psychologists, and other school resource experts to improve the quality of life in children with sickle cell disease. Disease management, including medication adherence to hydroxyurea or other medications, is indicated to prevent pain episodes that affect the quality of life (Thornburg et al., 2012; Wang et al., 2011). Patient management at school, including rest and hydration, may further prevent symptoms of pain, which may signal a sickle cell crisis. With school support, symptoms of depression and/or anxiety may be identified so a referral or intervention can be made. Students treated in a matter-of-fact and inclusive manner further enhances quality of life.

Conclusion

While clinicians focus primarily on the painful episodic exacerbations of sickle cell disease, which are often life-threatening, more attention needs to be directed at the assessment and management of psychosocial and school related factors. The Conceptual Model of Quality of Life (Ferrans et al, 2005) includes environmental, psychosocial, and biological factors that could give direction to the assessment of children and adolescents with sickle cell disease. During screening for developmental challenges and as part of preventive care, clinicians need to monitor academic performance, and explore barriers and challenges at school. Characteristics of the home and school environment must be considered, as well as family dynamics, which may affect the way a student views the chronic illness and quality of life. Strengthening the environmental support system necessary for a child with a chronic illness, such as sickle cell disease, may be a major factor in improving the quality of life and off-setting depression and anxiety.

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