Role of Spirituality in Patients with Sickle Cell Disease

Melanie Cooper-Effa, MD, MPH, Wayne Blount, MD, MPH, Nadine Kaslow, PhD, Richard Rothenberg, MD, MPH, and James Eckman, MD

Background: **Patients with sickle cell disease cope with their disease in various ways, such as psychological counseling, hypnosis, medication, and prayer. Spirituality is a coping mechanism in a variety of diseases. This study evaluates the role of spirituality in patients coping with the pain of sickle cell disease.**

Methods: **Seventy-one patients from the Georgia Sickle Cell Clinic completed a questionnaire addressing their ability to cope with the pain of sickle cell disease and their degree of spirituality. A descriptive cross-sectional design was used. Correlation and multiple regression analyses were calculated for the relation between coping with the pain of sickle cell disease and spirituality.**

Results: **The questionnaire provided several scales with high internal consistency for measuring spiritual well-being and its two components, existential well-being and religious well-being, that show a correlation between high levels of spirituality and life control. The study population exhibited high levels of spirituality and religiosity, but the influence of these feelings on coping with sickle cell disease was variable. Spiritual well-being was correlated with life-control but not with perceived pain severity.**

Conclusions: **Existential well-being was associated with general coping ability. Spiritual well-being is important for some patients who must cope with the pain of sickle cell disease. (J Am Board Fam Pract 2001;14:116–22.)**

Sickle cell disease is a hereditary hemoglobinopathy with a variable disease presentation and patient response pattern. It is categorized genotypically according to the predominance of hemoglobin S. This hemoglobinopathy can take on various forms, including hemoglobin SS, hemoglobin SC, and sickle β-thalassemia, with a clinical syndrome ranging from an asymptomatic course to a debilitating disease with increased mortality. Other characteristics include unpredictable, acute, and intermittent crises and chronic end-organ damage. Historically, these complications have led to shortened life spans. Improved medical care for these patients beginning in the 1970s, however, has helped them live longer, although patients with sickle cell disease still have shortened life spans when compared with the general population. The median age of survival in patients with sickle cell disease is now 42 years for men and 48 years for women.[1] Causes of early death in patients with sickle cell disease include renal insufficiency, acute painful crisis, acute anemic episodes, seizures, acute chest syndrome, pneumococcal septicemia, acute splenic sequestration, and aplastic crisis.[1,2]

Patients with sickle cell disease have an unpredictable course that can range from mild to severe disease. Severe disease has frequently been associated with a debilitating clinical course that decreases quality of life, causes frequent hospitalizations, and results in both work and school absenteeism from painful episodes, stroke, acute chest syndrome, sepsis, avascular necrosis of the femur, retinopathy, and osteomyelitis. Stroke is often recurrent and can lead to major functional deficits. Acute painful episodes and chronic pain are common; acute painful episodes increase with age and peak in the third and fourth decades of life.[2,3]

In an attempt to assess treatment and prognosis, investigators have described severity of sickle cell disease with little agreement on a single, validated method. Westerman et al[4] attempted to evaluate disease severity by painful episodes and classified severe painful episodes as those lasting 48 hours or more with irreversible tissue damage.

Submitted, revised, 1 May 2000.

From the Department of Family and Preventive Medicine (MCE, RR), the Department of Psychiatry (NK), and the Department of Medicine (JE), Emory University; Atlanta; and the Department of Family Medicine (WB), University of Tennessee, Memphis. Address reprint requests to Wayne Blount, MD, MPH, 1127 Union Ave, Memphis, TN 38104.

Much as sickle cell disease and its stresses are highly variable, so is the ability to cope with them. The severity of sickle cell disease, however, might not determine a patient's ability to cope. Coping with both sickle cell disease and its stresses can be a major struggle, resulting in lowered self-esteem and learned hopelessness.[2] Coping with disease is described by Lazarus and Folkman[5] as a twofold process of "adaptive functioning and regulation of emotional state." To enhance supportive disease treatment, elements that promote adjustment, health maintenance, and health education must be emphasized. Patients cope with disease in a variety of ways, for example, hypnosis, biofeedback, prayer, family support, drug use and abuse, behavior modification, and psychological counseling.[2,6–9]

Spirituality is another coping method in wellness and in a variety of diseases. Such spirituality is defined as the capacity to rise above life experiences, to celebrate life, and to experience joy.[9,10] Although closely related, spirituality and religiosity are considered distinct concepts. Spirituality applies to all mankind and involves the need for love, acceptance, forgiveness, and self-fulfillment, whereas religiosity applies to a specific religion or religious group.[11,12] Each concept plays a role in determining spiritual well-being.

Spiritual well-being has been described as a twofold state, including (1) a religious component (a positive specific relationship with God) and (2) a spiritual or existential component (a sense of life purpose and satisfaction).[13] There is a positive relation between spiritual well-being and psychological well-being, an inverse relation between existentialism and depression, and a direct association of spirituality with hope or future-oriented goal setting.[14] Spirituality can diminish depression, anxiety, and helplessness and can promote better mental health.[15]

Although regarded as an important element in the life process, there has been little emphasis on spirituality in medical practice, perhaps because it is considered a personal or nonscientific issue. Investigations of the affectation of spirituality and disease outcome, however, have documented a positive impact.[16–22] Researchers have found that spiritual health positively affects physical energy and improves health.[16] A positive correlation of lower blood pressure and religion-church attendance has been documented.[17] Also, religious faith has been shown to be an indicator of survival in patients undergoing elective cardiac surgery. There is a positive influence of prayer for patients in a coronary unit.[18,19] Spiritual well-being was also found to correlate with quality of life in patients with chronic disease, such as rheumatoid arthritis.[15] Spirituality has been highlighted as a supportive tool in health promotion with cancer cures, treatment of addiction, and other health issues.[20–22]

In the United States, sickle cell disease is found predominantly among the African-American population, a group that has traditionally regarded religion as an important aspect of their lives.[23] African-Americans are often frequent church attendees and have a high probability of being church members.[24] Although evaluated in other diseases, the role of spirituality has not been evaluated in patients with sickle cell disease. This study reports the role of spirituality in patients coping with the pain of sickle cell disease (both acute painful episodes and chronic pain). The study also tests the hypothesis that spiritual well-being and its components, religious well-being and existential well-being, enable patients to cope more effectively with the pain of sickle cell disease. (Henceforth, in this article pain means the pain of sickle cell disease and includes both acute painful episodes and chronic pain.)

Methods

Study Design

A descriptive cross-sectional design was used to examine the effect of spiritual well-being on life interference, pain severity, and life control. In October and November of 1997, patients older than 18 years attending the Georgia Sickle Cell Clinic in Atlanta were asked to complete a questionnaire that elicited information on demographics, coping skills, and spirituality. Patients were first contacted by mail or telephone. Questionnaires were completed at the clinic at a time when the patient was not in pain after informed consent was obtained. A $10 participation incentive was offered to reimburse respondents for the time required to complete the questionnaire. All 71 patients who were asked to complete the questionnaire participated. Medical records were reviewed to confirm sickle cell type and severity.

Key Measures

The dependent variable was coping with the pain of sickle cell disease, defined as leading an active life with good psychological adjustment. The major independent variable was spirituality pertaining to existential well-being and religious well-being. The Spiritual Well-Being Scale designed by Ellison[13] was used to measure spirituality. This instrument consists of a 20-item questionnaire with responses based on a six-point Likert scale, ranging from strongly agree to strongly disagree. The questionnaire is divided into two subscales: 10 items focus on life satisfaction and life direction (existential well-being), and 10 items focus on belief in God (religious well-being). Spiritual well-being was measured using scores from both subscales. The Spiritual Well-Being Scale has been validated by a number of investigators.[14,25–27] In a study by Carson et al,[25] a positive correlation between spiritual well-being and hopefulness occurred in a group of patients infected with the human immunodeficiency virus. Using factor analysis, internal consistency was reported to be 0.70 for total spiritual well-being and its components. Reliability was 0.94 for spiritual well-being, 0.92 for religious well-being, and 0.93 for existential well-being. Evidence for the construct validity of the Spiritual Well-Being Scale was described by Bufford et al,[27] who reported test-retest and internal descriptive data for several religious, student, and client groups.

Ability to cope with pain (dependent variable) was assessed with the West Haven-Yale Multidimensional Pain Inventory, which consists of 52 items in three parts.[28] The first two parts assess the patients' appraisal of pain (pain severity) and its impact on their lives, including their perception of psychosocial support (life control); the third part addresses the extent to which patients are able to participate in common daily activities (life interference). Each part is graded on a seven-point Likert scale. The construct validity of the West Haven-Yale Multidimensional Pain Inventory was calculated by its developers.[28] Reliability estimates for all scales are in the range of 0.7 to 0.9.

As noted, there is no universally accepted method validated for assessing severity of sickle cell disease. This study used a binary classification of severe and less severe. Patients were classified as having severe pain if they (1) were on chronic transfusion therapy, (2) had evidence of end-organ damage with complications, (3) had 12 or more medical visits for pain in the preceding 12 months, or (4) had required three or more hospitalizations per year. Patients with none of these characteristics were classified as having less severe pain.

Standard descriptive statistics, including mean, standard deviation, median, frequency counts, and percentages, were used to calculate sample demographics, clinical measures, and questionnaire results. Spearman rank order correlations compared the three pain-coping measures (pain severity, life control, life interference) with the overall Spiritual Well Being Scale and its subscales (existential well-being, religious well-being). The spiritual well-being measures were dichotomized into high and low categories to test for differences in pain measures using independent group *t*-tests and rank-sum statistics. Ordinary least squares regression evaluated the independent effect of the spirituality measures on the pain measures, controlling for age, sex, marital status, residence, hemoglobin type, and disease severity. Because of the study's exploratory nature, a stringent value tested for significance ($P < .01$).

Multiple regression analysis was used to control for disease severity and confounding variables, such as sex, age, marital status, and place of residence. This analysis also correlated coping ability with spirituality and disease severity, looking also at both subscales of the Spiritual Well-Being Scale. At a γ of 0.95, an α of 0.01, and an n of 71, the β was 0.95.

Results

In this older group of patients with sickle cell disease (mean age, 35.9 years), most had hemoglobin SS (65%) (Table 1). Sixty-one percent of the participants had severe disease, and 39% had less severe disease. For the major independent (spirituality) and dependent (coping) variables, Cronbach's alphas were greater than 0.8 for all measures, with the exception of pain severity and life control (0.644 and 0.679 respectively) (Table 2). Overall, this sample had high spirituality scores, with a mean score that was 80% of the maximum. With respect to our original question, the results shown in Tables 3 and 4 reveal the following: (1) spiritual well-being did not correlate with the level of interference with daily activities (life interferences), and (2) spiritual well-being did not correlate with the patients' appraisal of pain (pain severity), but (3) spiritual well-being did correlate with the patients'

Table 1. The Role of Spirituality in Patients with Sickle Cell Disease: Demographic and Medical Characteristics of Patients

Characteristic	Percent or Mean (n)
Sex	
Female	59 (42)
Male	41 (29)
Marital status	
Married	18 (13)
Single	64 (45)
Divorced	11 (8)
Separated	7 (5)
Hemoglobin type	
SS	65 (46)
SC	25 (18)
Other	10 (7)
12 or more medical visits for pain last year	
Yes	49 (34)
No	51 (36)
3 or more hospitalizations for sickle cell disease last year	
Yes	48 (34)
No	52 (37)
On chronic transfusion program	
Yes	9 (6)
No	91 (65)
Severe disease	
Yes	61 (43)
No	39 (28)
Age, years	35.9 (71)
Range, years (min/max)	18–63

perception of the disease's effect on their lives (life control). Of the two components of spiritual well-being, the existential component appeared to play a greater role than did the religious component.

Table 2. Descriptive Statistics for Spirituality and Pain Measures.*

Scale	Range (min/max)	Mean	Standard Deviation	Cronbach's Alpha
Spiritual well-being	48–120	100.72	15.90	0.882
Religious well-being	29–60	54.20	7.31	0.816
Existential well-being	19–60	46.52	10.55	0.829
Interference	0–6	3.27	1.53	0.863
Pain severity	0–6	3.35	1.53	0.644
Life control	0–6	4.08	1.56	0.679
Support	0–6	3.95	1.97	0.825

*Spirituality was measured using the Spiritual Well-Being Scale.[13] Pain was measured using the West Haven-Yale Multidimensional Pain Inventory.[28]

There was also a complex relation among disease severity, spirituality, and coping skills (Table 3). For example, life interference was more dependent on disease severity than on spirituality. In the presence of severe disease, high or low spirituality showed no significant difference in mean score (3.56 vs 3.54). For either level of spirituality, those with severe disease had a higher level of life interference than those with less severe disease. Life control, however, correlated more with the level of spirituality than with disease severity. At either level of disease severity, those with high spirituality scores had a higher score for life control than those with low spirituality scores.

Multiple regression analysis using life control as the dependent variable and controlling for age, sex, marital status, social support, hemoglobin type, and severity of disease suggests that existential well-being helps predict life control ($P = .0001$), accounting for 35% of the variance (Table 4). Religious well-being was also an independent predictor of life control ($P = .01$). When existential well-being was present, religious well-being had no effect. In similar regressions, using disease severity and life interference as the dependent variables, spirituality was not a significant predictor for either measure.

Discussion

In some studies, spiritual well-being is associated with improvement in disease outcome and quality of life.[14,15,18,19,22] How spiritual well-being might lead to these outcomes has been unclear. This study shows an association of spiritual well-being with better perception of life control for patients with sickle cell disease. While the association exists for both existential and religious well-being, the association with existential well-being is stronger. The association between disease severity and the major measures of coping (life interference, life control, and pain severity) is more complex. One can infer from this study, however, that the association found in past studies[14,15,18,19,22] between disease outcome and quality of life with spirituality might be influenced more by existential well-being. If a person is spiritually healthy existentially, that person has greater life satisfaction and life direction, as measured by this study. These two constructs are intuitively associated with quality of life and possibly even disease outcome via the psychoneuroendoimmunologic path.[15]

Table 3. The Role of Spirituality in Patients with Sickle Cell Disease: Mean Scores for Pain Scales by Spirituality and Disease Severity Classification.

Pain Scale	Spirituality, Disease Severity Classification*	Mean	Standard Deviation	*P* Value†
Interference	High spirituality, severe disease	3.6	1.3	.04
	High spirituality, less severe disease	2.8	1.7	
	Low spirituality, severe disease	3.5	1.8	
	Low spirituality, less severe disease	2.9	1.3	
Pain severity	High spirituality, severe disease	3.9	1.3	.02
	High spirituality, less severe disease	2.5	1.7	
	Low spirituality, severe disease	3.8	1.3	
	Low spirituality, less severe disease	2.9	1.4	
Life control	High spirituality, severe disease	4.6	1.3	.002
	High spirituality, less severe disease	4.7	1.0	
	Low spirituality, severe disease	3.1	1.7	
	Low spirituality, less severe disease	2.9	1.7	

*High spirituality defined as total score of 100 or more on the Ellison Spiritual Well-Being Scale.[13]
†Kruskal-Wallis alpha.

The population studied had a high degree of both religious and existential well-being, similar to previous observations about the African-American population.[23] The results of this study are in keeping with the body of evidence that has developed during the past decade associating spirituality in disease with an ability to tolerate pain. This study shows the importance of spiritual well-being in coping with the complications of sickle cell disease. Existential well-being (as a component of spiritual well-being) is associated with general coping ability. The effect of religious well-being on coping was not so strong.

Many clinicians might avoid issues of religiosity and spirituality because they view them as intensely personal, but this study suggests that these issues can be of therapeutic value in some patients.[29] The findings of this study support those of past studies showing a positive relation between spiritual well-being and psychological well-being, and between spirituality and hope.[14] Also supported are the study findings showing that spirituality can diminish anxiety and helplessness.[15] What is not clear is the method through which spiritual well-being increases life control. Is it by decreasing depression and anxiety, or by increasing hope, or vice versa? It could also be through a perception of greater social support; considering oneself to be spiritual might be correlated with a perception of a supportive network.

Table 4. The Role of Spirituality in Patients with Sickle Cell Disease: Multiple Regression of Existential Well-Being (EWB) and Religious Well-Being (RWB).

Dedependent Variable	Independent Variable	Adjusted R^2	*P* Value for F Statistic	F-Statistic	β Estimate	*P* Value for *t*-Statistic	*t*-Statistic
Interference	EWB only	0.06	.15	1.59	−0.03	.19	−1.33
	RWB only	0.04	.25	1.33	−0.00	.91	−0.12
Pain severity	EWB only	0.13	.03	2.36*	−0.02	.24	−1.20
	RWB only	0.13	.03	2.31*	−0.03	.29	−1.07
Life control	EWB only	0.36	.00	5.82†	0.09	.00	5.88†
	RWB only	0.10	.07	1.94	0.07	.01	2.65*
	EWB & RWB	0.35	.00	5.11†	—	—	—
	EWB	—	—	—	0.10	.00	4.97†
	RWB	—	—	—	0.01	.74	−0.34

Note: Controlling for age, sex, marital status, support, hemoglobin type, and disease severity.
*$P \leq .05$.
†$P \leq .0001$.

Clinicians should not expect spirituality to lower pain severity or the disease interference with daily activities. It would be reasonable, however, to expect some changes in a patient's overall perspective on how the disease affects his or her life. Clinicians might be able to assist their patients with a variety of coping strategies by inquiring about those that best suit the individual patient and encourage such healthy behaviors. If spirituality can help a patient cope, then clinicians should consider discussing the patient's spirituality. At least, the clinician can consider referral to a spiritual counselor. The clinician with limited time, but a desire to include the spiritual dimension with patients with sickle cell disease, can consider skipping questions about religiosity and spend time focusing on the questions of existential well-being. Suggested questions would be, "How satisfied or fulfilled are you with life?" "How good do you feel about your future?" "How much meaning does life have for you?" "Does your life have some real purpose?" "Do you know who you are and where you are going?." There might also be an existing need for religious counseling as indicated by client request.[30]

Although the instruments used have been validated in the past, there is a potential confounder in the assessment of disease severity used here. Three factors used in determining disease severity are primarily medical. The fourth factor, however, could be seen as determined more by a patient's perception of his or her illness and its severity (12 or more medical visits for pain). Even so, in this study there was no patient whose disease severity was determined only by that factor.

Conclusion

Spiritual well-being can help patients with sickle cell disease cope more effectively with the pain of sickle cell disease. This effect appears to be through a perception of more life control, measured in this study through the perception of social support. Considering oneself to be spiritual is correlated with perceiving oneself to have a supportive network. Spiritual well-being is not correlated with pain severity or life interference.

Further prospective studies involving interventional approaches to coping strategies and enhancement of spirituality are warranted. Man is a spiritual being. More effort should be placed on the development of that spirituality to enhance further wellness and quality of life.

The authors would like to thank Lewis L. Hsu, MD, PhD, Department of Pediatrics, Division of Hematology/Oncology and Bone Marrow Transplantation at Emory University School of Medicine for his contributions to this article.

References

1. Platt OS, Brambilla DJ, Rosse WF, et al. Mortality in sickle cell disease. Life expectancy and risk factors for early death. N Engl J Med 1994;330:1639–44.
2. Dover GJ, Vichinsky E, Graham SR, Eckman JR. Update in the treatment of sickle cell anemia: issues in supportive care and new strategies. In: Education Program of the American Society of Hematology. Orlando: National Institutes of Health, 1996: 21–32.
3. Pain in sickle cell disease. N Engl J Med 1991;325: 1747–8.
4. Westerman MP, Bailey K, Freels S. Assessment of disease severity in patients with sickle cell disease. Paper presented at the 19th annual meeting of the National Sickle Cell Disease Program, 23–26 March 1994, New York.
5. Lazarus RS, Folkman S. Stress, appraisal, and coping. New York: Springer, 1984.
6. Gil KM, Abrams MR, Phillips G, Keefe FJ. Sickle cell disease pain: relation of coping strategies to adjustment. J Consult Clin Psychol 1989;57:725–31.
7. Thompson RJ Jr, Gil KM, Abrams MR, Phillips G. Stress, coping, and psychological adjustment of adults with sickle cell disease. J Consult Clin Psychol 1992;60:433–40.
8. Whitten CF, Bertly JF. Sickle cell disease. Ann New York Acad Sci 1989;565:104–6, 183–205.
9. Kuhn C. A spiritual inventory of the medically ill patient. Psychiatr Med 1988;6:97–9.
10. Vastyan EA. Spiritual aspects of the care of cancer patients. CA Cancer J Clin 1986;36:110–4.
11. Chapman CS. Spiritual health: a component missing from health promotion. Am J Health Promotion 1986;1:38–41.
12. Highfield MF, Cason C. Spiritual needs of patients: are they recognized? Cancer Nurs 1983;6:187–92.
13. Ellison CW. Spiritual well-being: conceptualization and measurement. J Psychol Theol II 1983;11:330–40.
14. Landis BJ. Uncertainty, spiritual well-being, and psychosocial adjustment to chronic illness. Issues Ment Health Nurs 1996;17:217–31.
15. Hays RZ. Spiritual well-being and wellness in rheumatoid arthritis [master's thesis]. Seattle, Pacific Lutheran University, 1996.
16. Tubesing DA. Wholistic health: a whole-person approach to primary health care. New York: Human Science Press, 1979.
17. Larson DB, Koenig HG, Kaplan BH, Greenberg RS, Logue E, Tyroler H. The impact of relation in men's blood pressure. J Relig Health 1989;28:263–78.
18. Oxman TE, Freeman DH, Namheitre ED. Lack of

social participation or religious strength or comfort as risk factors for death after cardiac surgery in elderly. Psychosom Med 1995;57:5–15.

19. Byrd RC. Positive therapeutic effects of intercessory prayer in a coronary care unit population. South Med J 1988;81:826–9.
20. Justice B. Who gets sick: how belief, moods, and thought affect your health. Los Angeles: J P Tarcher, 1988.
21. McDowell D, Galanter M, Goldfarb L, Lifshutz H. Spirituality and the treatment of the dually diagnosed: an investigation of patient and staff attitudes. J Addict Dis 1996;15:55–68.
22. Magana A, Clark NM. Examining a paradox: does religiosity contribute to positive birth outcomes in Mexican American populations? Health Educ Q 1995;22:96–109.
23. Lincoln CE, Mamiya LH. The Black church in the African-American experience. Durham, NC: Duke University Press, 1990.
24. Taylor RJ. Religious participation among elderly blacks. Gerontologist 1986;26: 630–6.
25. Carson VB, Soeken KL, Shanty J, Terry L. Hope and spiritual well-being: essentials for living with AIDS. Perspect Psychiatr Care 1990;26:28–34.
26. Ledbetter MF, Smith LA, Vosler-Hunter WL, Fischer JD. An evaluation of the research and clinical usefulness of the Spiritual Well-Being Scale. J Psychol Theol 1991;19:49–55.
27. Bufford RK, Paloutzian RF, Ellison CW. Norms for the Spiritual Well-Being Scale. J Psychol Theol 1991;19:56–70.
28. Kerns RD, Turk DC, Rudy TE. The West Haven-Yale Multidimensional Pain Inventory (WHYMPI). Pain 1985;23:345–56.
29. Koenig H, George LK, Siegler IC. The use of religion and other emotion-regulating coping strategies among older adults. Gerontologist 1988;28:303–10.
30. McCullough M, Worthington EL, Maxey J, Rachel KC. Gender in the context of supportive and challenging religious counseling intervention. J Counsel Psychol 1997;44:1–9.