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Health-Related Quality of Life and Predictive Role of Self-Efficacy in Iranian Patients with Sickle Cell Disease

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ABSTRACT

Background: Although several studies have been done on quality of life of patients with Sickle Cell Disease (SCD), there is little research on the correlation of health-related quality of life (HRQoL) with self-efficacy in these patients. We aimed to determine the association between HRQoL and self-efficacy in patients with SCD and to explore the role of self-efficacy and demographic-clinical variables in a sample of Iranian patients with SCD.

Methods: In this cross-sectional study, 97 SCD patients who had medical records in Shafa Hospital affiliated to the Ahvaz University of Medical Sciences were recruited. Data were collected using the Short-Form Health Survey SF-36 (RAND 36-item), the Sickle Cell Self-Efficacy Scale (SCSES) and a demographic-clinical information questionnaire during February to July 2013.

Results: The mean scores of physical and mental component summary of the SF-36 (PCS and MCS) was 45.58±19.94 and 48.1±19.63, respectively which were low in patients with SCD. Moreover, 50.5% of the patients reported a moderate level of self-efficacy (24.42±6.59). Regression models showed that self-efficacy was the most important predictor of the mental component summary (MCS) (β: 0.48, P=0.001). With a slight difference, it was the second strongest predictor of the physical component summary (PCS) (β: 0.28, P=0.003), after the variable of "renal disease history" in the context of SCD (β: -0.30, P=0.001). However, "blood transfusion history" was a common predictor for both the PCS (β: 0.20, P=0.03) and the MCS (β: 0.26, P=0.001) components of the HRQoL.

Conclusion: The results of this study can assist health policy makers and clinicians to plan holistic interventions by focusing on the level of self-efficacy in SCD patients.

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Introduction

Sickle cell disease (SCD) is a monogenic hereditary disease. It is one of the most common genetic disorders worldwide characterized by chronic hemolysis, vaso-occlusive crisis and systemic inflammation.[1,2] The incidence rate of SCD is over 300,000 new cases worldwide per year.[3] In Iran, SCD is found in the southern provinces, especially in Khuzestan. According to the latest statistics, there are about 500 patients with SCD registered in Khuzestan.[4]

SCD has a debilitating course and patients suffer from unpredictable chronic pain episodes which are one of

the most commonly reported complications by patients and leads to frequent referrals to the emergency room and hospital for medical care.[5, 6] The other significant complications of SCD include acute and chronic anemia, acute chest syndrome, impaired vision, stroke, gallstones, priapism, renal failure, splenic sequestration, bacterial septicemia and chronic organ damage which all could result in impaired functioning and social isolation particularly in adolescent and young adults.[7-9] Confronting the challenges of this chronic disease influence the health-related quality of life (HRQoL) of these patients compared with the general population. Such problems can lead to differences in physical, psychological and social functioning of patients with SCD and make their quality of life more vulnerable.[10, 11] In this study, HRQoL as an outcome has been implemented as the patients' perception of well-being, along with physical, psychological and social aspects of the disease, according to the World Health Organization definition.[12]

A review of literature shows that there are not much studies about the impact of the SCD and its complications on quality of life of the affected subjects.[11] Several studies have reported poor quality of life in SCD patients.[13-18] Numerous factors affect patients' HRQoL and make it subject to obvious fluctuations. The results of some studies have shown that there is a direct relationship between pain and HRQoL of the patients, so the patient's HRQoL significantly improved after using pain relievers.[17, 19] Hijmans and colleagues found a decline in HRQoL and its relationship with low socioeconomic status in the population with SCD.[20] Imhonde and co-workers reported a direct relationship between HRQoL of patients with SCD and their self-esteem and social support, but an inverse correlation with depression.[21] Moreover, Ziadni and colleagues found that HRQoL of adolescents with SCD was associated with their adaptive behavior. Thus, the higher levels of coping correlated with better quality of life.[22] Self-efficacy is recognized as one of the key factors that contribute to HRQoL of these patients. Self-efficacy is a person's belief in his/her ability to perform tasks that are related to the daily management of symptoms and illness and is the basis of the changes in the behavior in a motivational theory.[23] According to Bandura's cognitive social theory, the belief of a person in his/her own capabilities to overcome the specific challenges can lead the person to overcome obstacles and have a better feeling of satisfaction.[24] Recent findings have showed that SCD patients with higher self-efficacy have higher levels of HRQoL.[23] Furthermore, patients with a lower level of self-efficacy have reported more pain and more physical and psychological symptoms associated with SCD.[25, 26]

A review of previous studies shows that although several studies have been focused on quality of life of SCD patients abroad, little research has been done regarding the association between self-efficacy and quality of life in these patients particularly in Iran despite the high prevalence of the disease in Khuzestan province. In addition, knowledge in this area could help further establishment of social and health policies for strategic planning and preventive strategies, timely diagnosis and management of problems pertaining to various aspects of life and thus health promotion in SCD patients. The aims of this study were to determine HRQoL and self-efficacy of patients with SCD and to explore the predictive role of self-efficacy and demographic-clinical variables in a sample of Iranian patients with SCD in Ahvaz city, center of Khuzestan.

Materials and Methods

Participants and Data Collection

All patients who were regularly visited at the hemoglobinopathy department and outpatient clinic of Shafa Hospital affiliated to Ahvaz University of Medical Sciences, were asked to participate in this study based on inclusion criteria. Totally, 97 patients with SCD over 16 years were enrolled in the study by a consecutive sampling between February to July 2013. Inclusion criteria in our study were: 1) patients with Iranian nationality older than 16 years old, 2) diagnosis of sickle cell disease or sickle thalassemia in the medical records and 3) ability to read and write in the Persian language and to answer to the questionnaires. Pain crisis episode at the time of the assessment of the patients was considered as exclusion criteria.

This study was approved by the Research Ethics Committee of the Shahid Beheshti University of Medical Sciences (code number: IR.SBMU.PHNM.1395.450) and Ahvaz Jundishapur University of Medical Sciences (code number: IR.AJUMS.REC.1395.730). All selected subjects and their guardians were informed about the aims of the study. Those who participated in the study signed the informed consent form. Data were collected through three questionnaires by a trained registered nurse in the hospital. Patients answered to the questionnaires in a private room.

Instruments

Demographic-Clinical Information Questionnaire: The demographic-clinical information questionnaire consisted of personal and clinical data, i.e., age, sex, marital status, level of education, job status, sickle cell disease genotype, treatment with hydroxyurea and patient history regarding blood transfusion, splenectomy, heart, renal, bone and joint diseases.

Short-Form Health Survey SF-36 (the RAND 36-item): The Short-Form Health Survey SF-36 (the RAND 36-item) instrument was used for assessing HRQoL. It is comprised of eight domains: physical functioning (10 items), role limitations due to physical health problems (4 items), role limitations due to emotional problems (3 items), energy/fatigue (4 items), emotional well-being (5 items), social functioning (2 items), pain (2 items) and general health (5 items). Scores range from 0 to 100 with 100 representing the best level of functioning or well-being.[27-29] Four subscales related to physical health were represented by the "Physical Component Summary (PCS)", i.e., physical functioning and role limitations due to physical problems, pain, and general health. Four

subscales related to mental health were represented by the "Mental Component Summary (MCS)", i.e., social functioning, role limitations due to emotional problems energy, and emotional well-being.[27, 30] The instrument was previously translated and validated into the Persian language and cross-culturally adapted for Iranian population.[27] In the present study, the reliability of the RAND 36-item using Cronbach's alpha coefficient was estimated to be in range of 0.70-0.83 in all subscales.

Sickle Cell Self-Efficacy Scale (SCSES): The Sickle Cell Self Efficacy Scale (SCSES) is a 9-item disease-specific instrument. This scale was designed and developed by Edwards and co-workers.[31] Higher scores indicate greater self-efficacy for coping with SCD. The SCSES uses a 5-point Likert scale ranging from 1 (not at all) to 5 (completely sure). The minimum score is 9 and the maximum score is 45, a higher score indicating higher self-efficacy. Defined cut-off points are as follows: A score of 9 - 20 is regarded as low, 21 - 32 as moderate, and 33-45 as high self-efficacy. The SCSES is a reliable scale with a Cronbach's alpha coefficient of 0.89, estimated by Edwards and colleagues in 2000.[31] Also, this scale was translated and validated into the Persian language (Cronbach's alpha coefficient of 0.74).[32]

Table 1: Demographic and clinical characteristics of patients with SCD based on the PCS, the MCS and the SCSES (n=97)

Variables		Number (%)	PCS Mean±SD	MCS Mean±SD	SCSES Mean±SD
Age	<24	53(54.6)	48.79±19.79	50.05±17.75	24.56±6.68
	≤25	44(45.4)	41.72±19.65	45.75±21.67	24.24±6.56
	P value		0.080	0.200	0.8
Sex	Male	63(64.9)	43.73±20.56	52.91±19.21	25.79±7.25
	Female	34(35.1)	46.58±19.69	45.50±19.52	23.68±6.14
	P value		0.500	0.070	0.1
Marital status	Single	67(68.0)	46.73±20.80	48.57±19.47	24.35±6.85
	Married	30(31.0)	43.02±17.92	47.03±20.29	24.56±6.08
	P value		0.400	0.700	0.8
Job status	Unemployed	55(56.7)	44.53±21.15	45.56±19.81	23.76±7.13
	Employed	22(22.6)	56.74±16.69	56.74±16.69	25.36±6.04
	Student	20(20.6)	48.39±1566	50.94±19.73	25.20±5.64
	P value		0.360	0.040	0.53
Educational level	≤High school Diploma	86 (88.7)	45.81±20.60	47.87±18.64	23.93±6.25
	University	11 (11.3)	43.81±14.30	49.86±27.24	28.27±8.14
	P value		0.600	0.800	0.03
Sickle cell disease genotype	HbSS*	63(64.9)	44.38±20.01	52.01±18.6	24.61±6.44
	S-ß-Thal **	34(35.1)	47.81±19.92	40.84±19.69	24.05±6.94
	P value		0.400	0.007	0.6
Blood transfusion	Yes	78(80.4)	42.78±19.91	44.35±18.53	23.71±6.38
	No	19(19.6)	57.07±15.83	63.46±16.67	27.31±6.84
	P value		0.005	0.001	0.03
Hydroxyurea consumption	Yes	52(53.6)	43.65±19.12	42.82±18.81	23.50±6.45
	No	45(46.4)	47.81±20.84	54.20±18.97	25.48±6.67
	P value		0.300	0.004	0.1
Iron-chelating therapy	Yes	60(61.9)	45.88±20.93	43.98±19.79	23.4±6.05
	No	37(38.1)	45.10±18.48	54.77±17.66	26.08±7.16
	P value		0.800	0.008	0.05
Splenectomy	Yes	48(49.5)	45.25±22.09	43.44±18.23	23.54±6.09
	No	49(50.5)	45.91±17.80	52.66±20.06	25.27±7
	P value		0.800	0.020	0.1
Bone and joint disease	Yes	82(84.5)	44.21±20.04	47.02±20.01	24.1±6.73
	No	15(15.5)	53.08±18.17	53.98±16.81	26.13±5.66
	P value		0.100	0.200	0.2
Heart disease	Yes	14)14.4)	29.45±13.95	34.36±11.78	19.14±4.92
	No	83(85.6)	48.30±19.56	50.41±19.79	25.31±6.44
	P value		0.001	0.001	0.001
Renal disease	Yes	17(17.5)	29.12±13.43	40.84±19.71	21.35±5.62
	No	80(82.5)	49.08±19.39	49.64±19.39	25.07±6.63
	P value		<0.001	0.090	0.03
Acute chest syndrome	Yes	67(69.1)	45.08±21.32	46.37±20.41	23.88±7.05
	No	30(30.9)	46.71±16.72	51.96±17.48	25.63±5.34
	P value		0.600	0.100	0.1

*Sickle Cell Anemia; **Sickle Beta Thalassemia

Data Analysis

Data analysis was conducted using the SPSS version 19. Descriptive statistics (frequency, mean and standard deviation) were calculated. Independent t-test and one-way ANOVA tests were used to compare the mean score of the PCS and MCS and self-efficacy in terms of dichotomous demographic and clinical variables. Then, significant demographic and clinical variables were entered into the regression models as independent variables. The relationship between outcome variables (the PCS, the MCS and SCSES) were estimated in a correlation matrix by Pearson correlation coefficients. Multiple linear regression analyses by stepwise method were conducted to determine the predictors of HRQoL in SCD patients.

Results

The results showed that the mean age of participants was 25.57±7.09 years. More than half of the patients were females (64.9%) and single (68%). The most common reported complications among SCD patients were Joint disease (84.5%) and acute chest syndrome (69.1%), respectively (Table 1). Demographic and clinical characteristics of the patients according to HRQoL (the PCS and the MCS) and self-efficacy mean scores are presented in Table 1. The results showed that the PCS mean score was significantly different between classifications of three variables (blood transfusion, heart disease, renal disease) The MCS mean score was also significantly different between classifications of seven variables (job status, SCD genotype, blood transfusion, hydroxyurea consumption, Iron-chelating therapy, Splenectomy, heart disease). Based on these results, significant variables were entered into the regression models as independent variables. The mean score of the RAND 36-item (eight subscales and two summaries) and the SCSES in SCD patients are shown in Table 2. The mean score of the PCS, MCS and SCSES were 45.58±19.94, 48.1±19.63 and 24.42±6.59, respectively. Mean score of self-efficacy in patients indicates that 50.5% of them reported a moderate level of self-efficacy, while only 13.4% of patients showed a high level of self-efficacy. Patients with education at the level of the university showed a significantly higher level of self-efficacy compared to those who had lower education (P=0.03).

Table 2: Descriptive data of the RAND SF-36, two components of the PCS and the MSC and the SCSES in patients with SCD (n=97)

Scales of the SF-36	Mean±SD
Physical functioning	63.81±21.61
Role limitations due to physical health	29.41±35.51
Role limitations due to emotional problems	33.33±40.25
Energy/Fatigue	46.86±18.82
Emotional well-being	54.72±20.26
Social functioning	57.47±24.78
Pain	48.48±28.51
General health	40.62±20.14
PCS	45.58±19.94
MCS	48.1±19.63
SCSES	24.42±6.59

Before regression analysis, the relationship between the outcome variables of the study were evaluated through a correlation matrix. A significant positive correlation was found between PCS (r: 0.39, P: 0.001) and MCS (r: 0.56, P: 0.001) with SCSES. Based on the previous studies, it was assumed that self-efficacy and demographic-clinical variables can predict HRQoL components (PCS and MCS) in patients with SCD in the regression models (Table 3). The results of regression models showed that self-efficacy was the most important predictor of the MCS (β: 048, p: 0.000). But for the PCS, it was the second strongest predictor (β: 0.28, p: 0.003), after "renal disease history" as one of the side effects of the disease (β: -0.30, p: 0.001). Self-efficacy together with clinical variables explained 26% and 44% of the variance in the PCS and the MCS, respectively.

Discussion

Based on our knowledge, research on the HRQoL and self-efficacy together among patients with SCD in Iran has not received enough attention. The current study showed that the mean score of physical and mental components of HRQoL for SCD patients were lower than the standardized mean score of 50 (SD:10) for population norms on the SF-36.[33] HRQoL of the patients was very

Table 3: Summery results of multiple linear regression analysis by stepwise model with the PCS and the MCS as dependent variables

Dependent variables	Summery model	Predictor	B	SE	β	t	P
PCS	R=0.53	Constant	0.157	0.082	-	1.92	0.05
	R^2=0.28						
	Adj.R^2=0.26	Self-efficacy	0.009	0.003	0.283	3.08	0.003
	F=12.23	Renal disease	-0.158	0.047	-0.304	-3.37	0.001
	df1=3	Blood transfusion	0.98	0.045	0.197	2.18	0.03
	df2=93						
	P<0.001						
MCS	R=0.68	Constant	0.189	0.094	-	2.011	0.04
	R^2=0.47						
	Adj.R^2=0.44	Self -efficacy	0.014	0.002	0.478	6.08	0.001
	F=20.44	Blood transfusion	0.126	0.038	0.256	3.28	0.001
	df1=4	Sickle cell disease genotype	-0.081	0.032	-0.197	-2.53	0.01
	df2=92	Hydroxyurea	-0.063	0.031	-0.162	-2.06	0.04
	P<0.001						

low, especially in the subscales of role limitations-physical, role limitations-emotional and general health, respectively. Our findings are in line with previous studies which have also reported poor level of HRQoL in SCD patients.[11-15, 17, 18] These are not surprising results due to the disease and its complications such as unpredictable pain crisis episodes leading to frequent visits to the hospital, chronic damages to the organs, physical and mental dysfunction, social isolation, financial and employment problems as well as increasing mortality risk.[5-11]

Moreover, around half of the patients in our study reported a moderate level of self-efficacy. These results are consistent with the previous studies.[34] However, there are different studies which have reported a low level of self-efficacy in the SCD group of the patients.[23] It seems that the variation between the results is due to methodological differences among the studies.

The results of regression analysis in the last model showed that self-efficacy was the most important predictor of mental component of HRQoL. Self-efficacy with a slight difference was the second predictor of physical component of HRQoL, after "renal disease history" in the context of SCD. This is in line with earlier studies. They found that self-care management resources (e.g., self-efficacy, social support, assertiveness, and self-care ability) were positively associated with quality of life and positive health outcomes, when tailoring for sociodemographic variables.[10, 35, 36-39] While, "blood transfusion history" was a common predictor for physical and mental component of HRQoL in our study, "genotype of the disease" and "Hydroxyurea consumption" were the next predictors for mental component of HRQoL. Based on previous studies, disease severity, including the occurrence of disease related complications, comorbid conditions, and treatment modalities (e.g., use of hydroxyurea and blood transfusion) have been described as determinants of HRQOL.[10, 35, 36-39] In these studies, more severe disease and greater healthcare utilization were associated with lower HRQOL in SCD patients.

In the present study, patients with Hb SS genotype reported better mental component of HRQoL in comparison to Sickle/Beta Thalassemia. In Mastandrea et al.'s study, the more severe genotypes of the disease (SS and S/β thal) reported worse HRQoL in two subscales of the physical component of HRQoL (general health and role limitations-physical), but not in the mental component. Appropriate management of the treatment and patients' adherence to it, in addition to patient access to more resources can influence mental HRQoL.[40]

In our study, patients with blood transfusion history showed a lower HRQoL in both the physical and mental components which was consistent with findings of previous studies.[14, 35, 41] In contrast, Beverung et al. found that patients who had history of chronic blood transfusion reported better HRQoL. Blood transfusion is a medical measure for reduction of many risks and complications of SCD patients, such as hemoglobin S concentration, pain, acute chest syndrome and primary stroke.[42] On the other hand, iron overload due to frequent blood transfusions could cause complications like iron accumulation in the liver, heart, skin, and other tissues resulting in serious tissue damage.[43]

Treated patients with hydroxyurea, reported lower level of HRQoL in the mental component. Recurrent pain is the main indication for hydroxyurea prescription and optimizing use of hydroxyurea throughout the lifespan is essential. In the contrary, some studies showed that SCD patients who take hydroxyurea reported a better HRQOL.[44-46] It should be noted that the mean age of the patients in our study was higher than reported in other studies. Badawy et al. found that a number of participants had challenges with hydroxyurea adherence which can be related to negative beliefs and its side effects.[7] This contradictory results are challenging and require further investigation. In summary, self-efficacy together with clinical variables explained 26% and 44% of the variances in the physical and mental components of HRQoL, respectively.

In our primary analysis with demographic variables, we found no statistically significant difference for HRQoL between the groups in terms of age, sex and marital status. While a previous study by Adzika et al. revealed that sociodemographic characteristics of SCD patients had a major role in their HRQoL.[41] Dampier et al. found that all the SF-36 subscales scores reduced along with increasing age.[35]

There are several limitations in this study that make it necessary to interpret the study findings with caution. First, the sample size was small which limits the generalizability of the findings. Secondly, the cross-sectional design of the study did not allow for measurement of the variables over time. Finally, self-report bias may have been introduced by the participants in response to the items of the questionnaires.

Conclusion

In this study the HRQoL of SCD patients in the physical and mental components of HRQoL was low. Also, around half of the patients reported a moderate level of self-efficacy. Self-efficacy together with several clinical variables explained 26% and 44% of the variances in the physical and mental components. Self-efficacy was the most important predictor of the mental component of HRQoL. With a small alteration, it was the second strongest predictor of the physical component of HRQoL, only lower than "renal disease history" in the context of SCD. These results can assist health policy makers and clinicians to plan holistic interventions by focusing on the level of self-efficacy in SCD patients.

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Conflict of Interest: None declared.

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