Saudi Journal of Medicine
Scholars Middle East Publishers
Dubai, United Arab Emirates
Website: http://scholarsmepub.com/

ISSN 2518-3389 (Print)
ISSN 2518-3397 (Online)
DOI: 10.36348/sjm.2017.v02i01.002

Original Research Article

Health-Related Quality of Life in Adults with Sickle Cell Disease in the Kingdom of Bahrain (FPRP)

Eman Ali, M.B., B.Ch., BAO, ABFM[1], Fatima F. AlHalwachi, M.D., ABFM[1], Fatima Khalil, M.B., B.Ch., BAO, ABFM[1], Zahra M. Hejab, MBBS, ABFM[1], Zainab A. AlMoalem, M.B., B.Ch., BAO, AFBM[1], Mohamed A. Mandil MBBS, AUB (Family Practice) M.Med.Sci (UK).[2]

[1]Family Physician, primary care, Ministry of health, Kingdom of Bahrain
[2]Consultant Family Physician, Private General Practice, Supervisor for residents in FPRP, Kingdom of Bahrain

***Corresponding Author:**
Eman Ali
Email: EAli14@health.gov.bh

Abstract: Sickle cell disease is one of the commonest reasons for hospital admissions in Bahrain and it considered one of the most significant causes of morbidity and mortality. However, HRQOL in SCD patients is not well studied in this part of the world. SF 36 questionnaire was administered to a total of 1886 participants, through manual and online methods in Salmanya Medical comlex and health centers outpatient's clinics, general society, and through SCD society, out of which 168 were excluded. Of the participants, 420 (24.4%) had SCD, 607 (35.3%) had chronic disease and 691 (40.2%) were disease free. SCD respondents scored significantly lower than the other two groups in HRQOL in all domains of SF-36, with the following means in role-physical 34.1 (SD 36.1), role-emotional 40.6 (SD 41.0) and general health 44.7 (SD 17.7) affected mostly $P \leq 0.001$. Adults with SCD in Bahrain showed significant impairment in all domains of HRQOL when compared to the general population. We would recommend conducting further researches to evaluate the factors that may influence the HRQOL in SCD patients**.**

Keywords: Health related quality of life, HRQOL, Sickle cell disease, SCD, Adult, Bahrain.

BACKGROUND

Sickle cell disease (SCD) is an autosomal recessive disease that is expressed as sickle cell anemia, sickle cell thalassemia and sickle hemoglobin C disease. Most patients in the gulf region have the Arab/Indian haplotype, which is a mild form of the disease with elevated HbF [1]. SCD affects around 300,000 people around the world [2]. It is considered one of the most common haemoglobinopathies in the kingdom of Bahrain with an incidence of 21 per thousand among newborns in 1984. However, it has dropped to 4 per thousand in 2013 [3].

Sickle cell disease is one of the commonest reasons for hospital admissions in Bahrain and has the highest rate for multiple readmissions [4]. The mortality rate among SCD in ICU in SMC is 12.7% [4]. Moreover, SCD was accounted for 30% of the maternal deaths between 1997 and 2012 in Bahrain [5].

The disease is characterized by chronic hemolytic anemia, increased susceptibility to infection, extensive organ damage, intermittent pain episodes, chronic pain [6] cardiovascular collapse and sudden death [4]. Moreover; it has been found that as the number of pain crises increases the mortality risk increases [7].

In addition to the severe physical symptoms and pain, psychological and social concerns associated with the illness have a major impact on quality of life for sickle cell patients [6].

Researchers have studied the impact of the chronic diseases through measuring the Health-Related Quality of Life (HRQOL) which has been shown to be a strong predictor of outcomes such as morbidity and mortality [8]. HRQOL can be defined as the individual's own subjective perception of aspects of life directly related to the state of health, including the satisfaction and wellbeing of an individual as related to the physical, psychological, social, economic and spiritual domains of his/her state of health [9].

Furthermore, quality of life measures are used in evaluating the efficiency of medical care [10], to improve patients-provider communication and to create a more patient – centered environment [8].

Increased life expectancy due to recent medical advances has raised the need to understand the quality

of life in patients with SCD and identify factors predicting disease adaptation [10].

A cohort study conducted on 1046 patients showed a substantial impairment in all subscales of HRQOL in adults with SCD except mental health, and suggests that more effective treatment of persistent pain and depression would provide the largest benefit [11]. Another study done in UK showed a decline in HRQOL in SCD individuals as a consequence of negative mood, daily activity and role limitations, and neurocognitive impairment [12].

The Genotype did not influence HRQOL except for vitality [13]. However, it was significantly decreased as pain level increased [13] and increasing age [11]. A Brazilian research has reported that the adult patients exhibited greater impairments in the functional capacity, mental health, social function, and role-emotional domains of HRQOL compared to the adolescents [9].

It has been showed that social support significantly correlates with QOL which will decrease feelings of isolation and improve social networks and social support [14], this is supported by the findings of qualitative study conducted in United Kingdom (UK), where participants spoke of the importance of having social support in coping with SCD [15].

However, HRQOL in SCD is not well studied in this part of the world, only one regional study was found which was conducted in Kingdom of Saudi Arabia (KSA) in 2011 and concluded that Saudi adolescents with SCD experienced deterioration among all domains of HRQOL especially physical [10]. To our knowledge, no study has been done in Bahrain to assess the HRQOL for individuals with SCD.

This study will be conducted to assess the impairment of the different domains of HRQOL among Bahraini adults with SCD compared to adults SCD and to define the relationship between socio-demographic variables and the severity of SCD with the degree of impairment in HRQOL. If these variables (socio-demographic and severity of SCD) are crucial in determining HRQOL among adults with SCD, thus by controlling these factors a better quality of life may be achieved with less burden on healthcare services.

METHODOLOGY

Aim

To improve the quality of life among adults with sickle cell disease in the Kingdom of Bahrain

Objectives

- To assess the major domains of quality of life.
- To identify factors that may be associated with poor QOL.
- To assess the QOL in relation to the severity of the SCD disease.

Study design

Cross sectional study

Sampling

This research included three groups; (1) adults with SCD, (2) adults with haemoglobinopathies other than SCD, including thalassemia, and decreased G6PD or chronic non-communicable disease like HTN, and DM and will be subsequently referred to as chronic disease group and (3) healthy adults referred to as No disease group. Convenient samples were collected by two methods; (a) manual distribution of the questionnaire, and (b) filling an online questionnaire.

The three groups were collected from Salmaniya medical complex – outpatient clinics department (SMC-OPD), five health centers, SCD-society and general population. A Self- Administered questionnaire was manually distributed among the attendants of Outpatient Clinics in SMC and the following primary care centers: Naim, A'ali, Jidhafs, Budaiye, Sitra, Ahmed Ali Kanoo (A.A.K), Yousif A. Rahman Engineer, and Bilad Al-Qadeem. Sitra, Jidhafs and Budaiya HCs were chosen due to the availability of SCD clinic. Naim, A'ali, Bilad Al-Qadeem and engineer HCs were chosen randomly.

SMC

SCD and non-SCD adults aged 18 years old and above were approached in the waiting area of the SMC-SCD evening clinic between February and June 2014. This clinic is a multidisciplinary clinic running twice weekly and serving approximately 17 SCD patients per day. The total number of those attendees who agreed to participate was around 50. A total of 16 SCD-adults were involved in the study from SMC Hematology morning clinic. Proper orientation about the study and its aim was carried out to the potential participants. A verbal consent was obtained before handing the questionnaire for those who agreed to participate in the study. Patients were then asked to complete all three sections of the questionnaire including the SCD related data section if they have SCD, and to skip it if they don't have SCD. Assistance was offered and provided by investigators when necessary.

Attendants of other SMC-OPD clinics were also approached in the waiting area, including: Cardiology, Medical, Neurology, Pediatric, and Surgical clinics. Similar approach described above was applied.

Health centers

Patients attending the above-mentioned health centers were approached while waiting for their

appointments including SCD and non-SCD individuals. SCD patients attending SCD clinic in Jidhafs HC were approached as well, and majority of them agreed to participate after understanding the potential benefits of the upcoming research. The number of attendants of HC's SCD clinic was small and majority was of pediatric age group. A small number of SCD patients were recruited from the treatment rooms of Jidhafs, Engineer, and Bilad Al-Qadeem HCs.

SCD-society and general population

Close contacts and friends of the researchers including SCD patients were asked to fill the questionnaire manually after a proper introduction of the research.

Survey monkey was used to generate an online questionnaire, which was distributed using SMS messages to the general population and through SCD-society to their mobile data base. A mobile number and e-mail address was provided for the online group, for any inquiries regarding the questionnaire. There were two online questionnaires, the first one was distributed through SCD-society only, and changed later on due to an error in data entry in the demographic section. The error was corrected in the second version of the questionnaire was distributed both to the general population and SCD-society mobile data base.

Measurements

Instrument

The SF-36 is a well-known generic health-related quality of life (HRQOL) measure that is widely used in various chronic conditions and populations [18]. The questionnaire addresses general health notions that are not specific in regards to age, disease or treatment group. SF-36 is a psychometrically sound, reliable and valid measure in many populations and is more responsive to clinical improvement than other instruments.

The SF-36 is suitable to persons age 14 and older. The SF-36 questionnaire can be self-completed or administered in person or over the telephone by a trained interviewer. It is considered simple to administer and takes less than 10 minutes to complete.

The original SF-36 came out from the Medical Outcome Study (MOS), done by the RAND Corporation in the United State of America and it has been translated to several different languages including Arabic. The psychometric testing for the SF-36 Arabic version approved its reliability and validity across different Arabian population and cultures [16, 17]. The median internal consistency reliability of Arabic version of SF-36 exceeded 0.70 for all scales except for the general health perception scale (median alpha = 0.59).

The SF-36 consists of 36 questions that are clustered to yield eight subscales, each evaluating a different domain of HRQOL a. Physical functioning 10 items, Role limitation due to physical health 4 items, Role limitation due to emotional problems 3items, Energy/fatigue 4 items, Emotional wellbeing 5 items, Social functioning 2 items, Pain 2 items and General Health 5 items. These subscales (domains) provide two summary scales, Physical Component Summary (PCS) and Mental Component Summary (MCS). A single item that provides an indication of perceived change in general health status over a one-year period is also included.

Scoring method

The SF-36 and RAND-36 include the same set of items that were developed in the MOS. They differ only in the scoring procedure of the general health and pain scales. RAND 36-Item Health Survey 1.0 was used in the research because it is simpler and more straightforward procedure [18].

It is a two-step process. First, pre-coded numeric values are recoded per the scoring key given in Table 1.(Appendix) Each item is scored on a 0 to 100 range so that the lowest and highest possible scores are set at 0 and 100, respectively. In step 2, items in the same scale are averaged together to create the 8 scale scores. Items that are left blank (missing data) are not considered when calculating the scale scores.

Data Analysis

The Statistical Package for the Social Sciences (SPSS-20) was used for the statistical analysis. Frequency and proportions was calculated for nominal and ordinal data. The collected sample was divided into 3 groups: SCD, respondents with chronic disease including other haemoglobinopathies and no disease. One way between groups ANOVA-test used for comparison of SF-36 subscales between the 3 groups, and the Chi-square test was used to compare the demographic data and 3 groups. A two way between groups ANOVA was used to explore the impact of demographic variables on physical and mental component summary. T-Test was used for the analysis of differences in SF-36 subscales among subgroups of SCD sample according to their presence of complications. Missing values were imputed for missing completely at random (MCAR) values.

Ethical consideration

The research proposal was submitted to the ethical committee, and an approval was obtained on Jan 2014. Following that permission letters were received from SMC and primary care administrations allowing for data collection within their facilities.

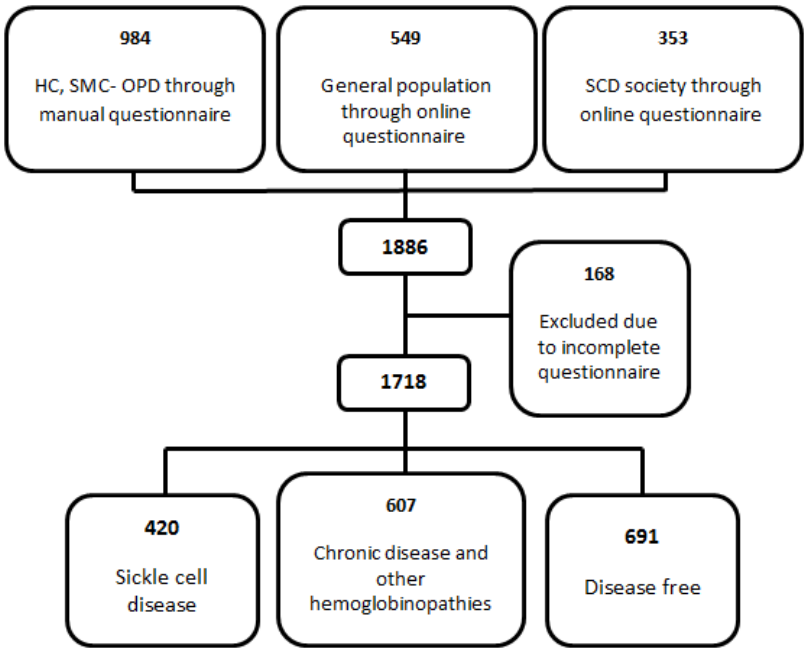

Fig- 1 : Flow chart summarizing total participants, excluded number and final sample included in the study

RESULTS

A total of 1886 participants were recruited to the study, of which 984 were collected manually from SMC and health centers outpatient's clinics and general society, 353 responded to online questionnaire through SCD society and 549 were through general population online distributed questionnaire (Fig 1).

Of the questionnaires returned, 168 questionnaires were not completed; consequently they were not included in the analysis.

Of the 1718 participants included in the analysis, 420 (24.4%) had SCD, 607 (35.3%) had chronic disease and 691 (40.2%) were disease free (Fig 1).

Socio-demographic data of the study sample

In the study sample, female respondents were over represented in all three groups (Table1). Majority of the respondents were aged between 18-30 (53.6%) years in SCD group and 325 (48.6%) no-disease group (Table 1). The mean age for all sample was 33.5 (SD 10.4) for SCD group, 30.7 (SD 8.6) for the chronic disease group 36.8 (SD 11.5) and for disease free group 32.4 (SD 9.7).

Majority of respondents in all groups had attained higher education level 54%, 60.3% and 62.2% in SCD, chronic disease and no disease group respectively (Table1).

Unemployment was observed to be higher among SCD group 56.7%, whereas it was lower in the other study groups 45.34%, and 45.3% in chronic disease and no disease group respectively (Table 1).

A higher proportion of the study sample were married in all groups, but with higher percentage 37% of single, divorced/widowed 5% individuals in adults with SCD compared to other groups (Table 1).

Although medium income was predominant in all three groups, low income was higher in SCD (15.5%) when compared to the other groups (Table 1).

Health related quality of life of the study sample:

A one-way between-group ANOVA was conducted to explore the impact of respondents with SCD, Chronic disease groups and respondents with no disease on mean score for the nine subscales of SF-36. There was a statistically significant difference in mean score between the groups $p < 0.001$. Respondents with SCD scored significantly lower than the other two groups in HRQOL L in all domains of SF-36. The effect size ranged from < 1% for emotional wellbeing and health change to large > 13.8% for physical functioning and general health (Table 2).

(Table 3) shows a two-way between-groups analysis of variance on mean Physical Health (PH) score between the groups SCD, Chronic disease and no disease group and demographic variables, age, gender, marital status, education, employment status and financial status. There was a significant interaction between the SCD group, chronic disease and no disease groups and gender $p < 0.000$ on PH mean score. Male respondents had lower mean score than females, but with small effect size 0.010. There was a significant main effect on PH mean score for SCD, Chronic disease group and no disease group $p < 0.000$. Respondents with SCD significantly scored lower than the other groups

against all domains and the effect size was moderate 6% to large 13.8%. Significant main effect was observed on PH mean score for age groups, education, marital status and financial status but the effect size was < 1%.

A two-way between-subject's ANOVA was conducted to explore the effect on mental health of respondents. Demographic data such as: gender, age-groups, education, employment, marital status had significant impact $p < 0.01$ on mean mental health score for participants with SCD. Being male, age group 31-40 years, below intermediate education level, unemployed, being divorced or widowed and low financial state. The effect size for all variables was small <1%.

The main impact of "groups" on mean mental health score was highly significant $p< 0.001$. Effect size was moderate 6%.

Respondents with SCD consistently and significantly scored lower than the groups with chronic disease and no disease.

Significant interaction was observed between "group", and gender and education $p< 0.01$ with small effect size, which indicates that the difference in mean mental health score between the groups (the effect of group) is different in males and females as well as in different education categories (Table 4).

Table 1: Socio-demographic data of the study sample values are number (%) N=1718

		SCD N=420		Chronic disease N=607		No disease N=691	
Patient Sex*	Male	198	(47.1)	250	(41.2)	211	(30.5)
	Female	222	(52.9)	355	(58.5)	480	(69.5)
Age	18 – 30	225	(53.6)	198	(32.6)	336	(48.6)
	31 – 40	135	(32.1)	205	(33.8)	227	(32.9)
	>40	60	(14.3)	204	(33.6)	128	(18.5)
Education*	Intermediate and less	42	(10.0)	45	(7.4)	34	(4.9)
	Secondary	149	(35.5)	194	(32.0)	226	(32.9)
	>College and Higher	227	(54.0)	366	(60.3)	427	(62.2)
Employment status*	No	238	(56.7)	275	(45.34)	313	(45.3)
	Yes	180	(42.9)	326	(53.7)	370	(53.5)
Marital status	Single	156	(37.1)	94	(15.5)	148	(21.4)
	Married	243	(57.9)	493	(81.2)	529	(76.6)
	Divorced OR Widowed	21	(5.0)	20	(3.3)	14	(2.0)
Financial status	High	124	(29.5)	196	(32.3)	231	(33.4)
	Medium	231	(55.0)	364	(60.0)	420	(60.8)
	Low	65	(15.5)	47	(7.7)	40	(5.8)

*Missing values

Table 2: One way between groups ANOVA SF-36 scores in adults with SCD, chronic disease and haemoglobinopathies and no disease "Mean (SD)"

	SCD		Chronic disease		No disease		ANOVA P value	η^2*
Physical functioning	56.6	(23.2)	68.3	(25.8)	73.0	(25.8)	**0.001**	**0.6**
Role-Physical	34.1	(36.1)	56.2	(39.5)	65.6	(36.1)	**0.001**	**0.09**
Role-Emotional	40.6	(41.0)	55.9	(40.7)	58.0	(39.0)	**0.001**	**0.02**
Energy Fatigue	45.4	(18.1)	50.7	(17.6)	53.7	(18.7)	**0.001**	**0.03**
Emotional Wellbeing	57.4	(19.9)	62.6	(17.9)	61.7	(18.1)	**0.001**	**0.01**
Social Functioning	60.0	(24.9)	69.7	(22.4)	72.7	(22.0)	**0.001**	**0.04**
Pain	51.6	(28.3)	67.9	(24.1)	73.3	(23.0)	**0.001**	**0.11**
General Health	44.7	(17.7)	59.1	(18.1)	64.7	(16.4)	**0.001**	**0.17**
Health change	52.7	(28.5)	57.1	(23.5)	58.3	(23.1)	**0.001**	**0.01**

η^2*Partial Eta squared

Cohen's criteria for effect size, small =0.01 (1%), medium=0.06 (6%), large = 0.138 (13.8%)

Table 3: Two-way between-groups ANOVA interactions on Physical Health score for disease groups and demographic variables. Mean (SD)

Physical Health					
	SCD	Chronic disease	No disease	P	η^{2}*
	Mean (SD)	Mean (SD)	Mean (SD)		
Gender					
Male	989.7 (374.6)	1392.5(418.7)	1515.7(376.0)	0.000	0.010
Female	1081.8(404.8)	1294.8(408.3)	1431.5(380.8)		
Marital status					
Single	1073.7(383.7)	1381.0(440.2)	1479.3(413.2)	0.99	0.000
Married	1027.9(401.2)	1331.2(408.0)	1455.2(371.0)		
Divorced/widowed	903.8(350.1)	1222.4(445.3)	1298.4(397.8)		
Age group					
18-30	1071.2(386.2)	1415.2(413.0)	1480.9(385.7)	0.36	0.003
31-40	996.9(381.4)	1320.2(418.2)	1446.0(368.1)		
>40	1020.2(423.2)	1284.7(399.4)	1465.2(375.8)		
Education					
<Intermediate	805.0(325.2)	1195.7(408.8)	1417.7(371.4)	0.11	0.004
Secondary	960.3(314.2)	1268.9(402.1)	1387.0(374.1)		
College/Univ.	1135.8(421.7)	1386.5(413.9)	1496.6(381.1)		
Employment					
No	1039.1(408.2)	1315.2(389.7)	1423.5(376.0)	0.5	0.001
Yes	1039.5(374.2)	1352.5(435.8)	1484.5(383.6)		
Financial status					
High	1136.8(414.9)	1369.0(440.6)	1513.4(367.8)	0.6	0.002
Medium	1035.7(379.9	1338.2(403.1)	1439.3(385.4)		
Low	861.8(334.5)	1179.4(365.8)	1341.5(372.7)		

*Partial Eta squared

Cohen's criteria for effect size, small =0.01 (1%), medium=0.06 (6%), large = 0.138 (13.8%)

Table 4: Results of two-way between- groups ANOVA for disease groups and demographic variables on Mental Health score mean (SD). N=1718

Mental Health					
	SCD N= 416	Chronic disease N=598	No disease N=690	P	η^{2*}
	Mean (SD)	Mean (SD)	Mean (SD)		
Gender					
Male	714.4(264.1)	915.7(239.10	922.8(241.8)	0.000	0.011
Female	735.8(274.5)	799.7(250.1)	838.6(253.4)		
Marital status					
Single	739.0(273.2)	825.9(255.6)	844.7(250.2)	0.228	0.003
Married	722.7(264.2)	853.2(250.7)	874.9(251.7)		
Divorced/widowed	664.8(311.2)	811.3(261.0)	665.7(237.2)		
Age					
18-30	718.4(269.3)	842.2(239.3)	842.1(264.9)	0.455	0.002
31-40	702.8(276.1)	813.8(257.9)	865.6(260.0)		
>40	804.0(251.2)	894.6(247.6)	952.4(229.0)		
Education					
<Intermediate	658.5(229.0)	798.0(229.5)	875.3(247.3)	0.018	0.007
Secondary	675.1(243.3)	845.4(231.8)	867.7(255.0)		
College/Univ.	772.3(285.1)	853.0(263.4)	861.6(253.0)		
Employment status					
No	719.2(273.0)	816.8(253.8)	844.6(256.6)	0.5	0.001
Yes	736.5(265.6)	872.5(247.9)	879.9(249.8)		
Income					
High	785.4(247.7)	873.3(260.8)	908.6(251.5)	0.671	0.001
Medium	724.3(277.5)	842.6(246.7)	848.1(250.7)		
Low	618.3(249.3)	782.7(245.8)	775.2(228.1)		

η^{2}*Partial Eta squared

Cohen's criteria for effect size, small =0.01 (1%), medium=0.06 (6%), large = 0.138 (13.8%)

Clinical characteristics of the adults with sickle cell disease:

From SCD sample, 315 (72.3 %) had visited the A/E department during the last 6 months due to SCD crises between 1 to > 4 times. Moreover, 322 (74%) were admitted to the hospital during the past year between 1 to > 6times and 129 (60.4%) were admitted at least once to >4 times in their entire life to the intensive care unit due to SCD complications. Only 75 (17.6%) from the SCD group reported using hydroxyurea (Table 5).

Majority of SCD participants 340 (81.7%), enrolled in the study experienced disease related complications of which bones and joints were the highest reported organs affected 263 (63.2%) followed by splenic complications 102 (24.5%) (Table 5).

Table 5: Clinical characteristics of the adults with SCD values are number (%)

Visit to A/E department	None	121	(27.8%)
	1-2	144	(33.0%)
	3-4	101	(23.2%)
	>4	70	(16.1%)
Admission to hospital	None	113	(26.0%)
	1-3	171	(39.3%)
	4-6	73	(16.8%)
	>6	78	(17.9%)
ICU admissions	0	85	(39.7%)
	1-2	90	(42.1%)
	3-4	29	(13.6%)
	>4	10	(4.7%)
Use of Hydroxyurea	Yes	75	(17.6%)
	No	350	(82.4%)
SCD Complications			
	Yes	340	(81.7%)
	No	76	(18.3%)
Lungs		34	(8.2%)
Bones and joints		263	(63.2%)
Kidneys		24	(5.8%)
Spleen		102	(24.5%)
Brain		10	(2.4%)

Health related quality of life in adults with sickle cell disease in relation to disease complication:

SCD individuals with disease related complications showed significant impairment in all domains of HRQoL, scoring the lowest in role-physical, role-emotional and general health compared to those without complications p< 0.001 (Table 6).

Table 6: SF-36 scores in adults with SCD according to the presence of complications "Mean (SD)"

	Complications (n=340)		No complications (n=76)		T-Test P-Value
Physical functioning	53.27	(22.18)	69.57	(22.18)	0.000
Role-Physical	26.33	(32.83)	62.50	(38.72)	0.000
Role-Emotional	35.52	(40.10)	59.05	(43.28)	0.000
Energy/fatigue	43.07	(18.40)	52.77	(18.07)	0.000
Emotional Wellbeing	55.20	(20.28)	65.60	(19.63)	0.000
Social Functioning	55.67	(24.51)	76.81	(21.89)	0.000
Pain	46.13	(27.50)	70.00	(26.93)	0.000
General Health	42.40	(16.60)	53.14	(19.38)	0.000
Health change	50.22	(28.68)	61.84	(26.61)	0.001

DISCUSSION

Adults with SCD in our study showed significant impairment in all domains of HRQOL when compared to the rest of sample. Role-physical, role-emotion and general health scales were mostly affected compared to the other groups in the study.

Comparable to the Bahraini individuals, SCD Saudi's adolescents had similar deterioration in all domains of HRQOL subscales [10]. In UK, SCD adults also scored less than the general population in all domains of SF-36 scales [20]. Qualitative studies done by Thomas and Taylor's and Helen Cair on patient with SCD found that SCD significantly impacted all domains of functioning [15, 19].

This negative effect of the disease on quality of life was consistent with other studies done on patients with significant chronic conditions like patients with cystic fibrosis, bronchial asthma and those undergoing hemodialysis [13].

Pain is an important dimension in the HRQOL of patients with SCD.A study done in UK concluded patients with sickle cell with pain who are admitted to day care and through A&E seemed to have impaired HRQOL as a result of pain [20].

We observed that SCD respondents who were males, age between 31–40 years, intermediate and below educational level, unemployed, divorced or widow and those who reported low income were scoring lower in pain scale when compared to the other demographic data. *Anie et al* [12] also found a relationship between pain and some subscales of the SF-36 (physical and social functions, mental and general health).

Sickle cell disease respondents, with complications, scored lower than those without complications. This finding was consistent with a study done on Saudi adolescents [10].

Among the reported SCD complications in our sample, cerebrovascular events were the least common, however; it has been found to be the most disabling complication in SCD patients [12].

From SCD sample 72.4% visited A/E once or more, 16.1% attended more than 4 times during the last year. It has been found in a Bahraini study that SCD patients who frequently presented to A/E demonstrated significant reduction in daily life functions with negative cognitive behaviors toward community [21].

Two thirds of SCD respondents have been admitted to the ICU at least once. The mortality rate among SCD in ICU in SMC is 12.7%. Four significant predictors of SCD mortality in ICU were identified; patient's age, number of hospital admissions, length of stay in ICU, and patient's need of mechanical ventilation. Non-survivors were older than survivors, less frequently admitted to hospital, have shorter length of stay in ICU, and usually come to ICU on mechanical ventilation [2].

Hydroxyurea has a significant effect in reducing the morbidity and mortality of SCD [22-23]. It shortens the duration of hospitalization because of acute painful episodes and reducing the net amount of opioid utilization [23]. In addition, treatment of SS with HU improves some aspects of QOL in adult patients who already suffer from moderate-to-severe SS [24]. However, majority of our study sample were not using the drug. This reluctance in receiving treatment with hydroxyurea may be attributed to the known side effects of the drug. One of the most feared side effects among Bahraini with SCD is the effect on fertility and reproductive life of the drug users.

In our study, HRQOL of female with SCD was more adversely affected when compared to males. Similarly, a regional study in Saudi Arabia showed significantly lower scores in physical functioning, role of physical, bodily pain and general health domains among females [10]. However; our study showed that Bahraini females scored significantly better in role-physical, social functioning, pain and general health.

Quality of life was not affected by the state of unemployment which was reported by 56.8% of the SCD individuals. This was contrary to a Jamaican study which found that subjective wellbeing was compromised in the unemployed SCD adults compared to employed SCD adults [25]. However, burden of the disease especially on the role of physical and physical functioning, may be explained by the unpredictability of SCD attacks, difficulty getting and keeping a job, and increased absenteeism from work might be the cause behind this finding.

Despite the high unemployment percentage among SCD sample, 55% reported medium income. This could be a result of the personal perception to financial status. Social and family support may also attribute to this discrepancy as well. Bahraini SCD respondents with low income compared to higher income scored significantly lower in all domains of HRQOL. Correspondingly, Saudi youth with low socioeconomic standard were likely to have lower physical functioning scores [10]. Possible explanation to our findings might be that individuals with lower income may be exposed to higher level of anxiety and stressful events due to lack of adequate financial and psychological support.

Although 57.8% of the enrolled SCD adults were married, 37.1% were single which was the highest between the three groups. This difference might be attributed to social and cultural factors. Having SCD is

considered a stigma and an obstacle when it comes to marriage and establishing a family, due to fear of passing the disease to offspring [15], and of the burden of the disease itself on person and family. In addition, along with the psychological stress, SCD patients experience a higher financial burden especially during crises. In a study done in Nigeria, relatives of patients in SCD crisis perceived similar financial, family routine burdens and psychological distress scores as cancer patients [26].

Married SCD individuals scored slightly lower than singles/divorced in which social function was the only significantly affected domain. Interestingly this is against the Brazilian study that found Married SCD individuals scored higher in all HRQOL domains except pain [27].

In the absence of a universal cure in SCD, the primary aim of treatment is to reduce the impact of the disease, thus enhancing quality of life [20].

Physicians play an essential role in the treatment of SCD, and with current belief that SCD patients are drug dependents and the concerns regarding drug abuse, they tend to underestimate their pain. This can serve as a major barrier to providing them with optimal pain management. This provider bias may lead to reluctance by patients to seek medical attention [12]. Consequently, this can have a major influence in quality of life and psychological and emotional belief for sickle cell patients. A study done in USA revealed that a moderate proportion of providers in the acute setting have negative attitudes toward patients with SCD-related pain [27].

Health care providers with negative attitudes toward SCD individuals, and those who reported caring for more than one SCD patient per week were less likely to adhere to the most important aspect of high quality SCD pain management [28].

Nurses being the front line in patient management are expected to be knowledgeable of their patients' disease, however; 47% of staff nurses were found in a Bahraini study to have significant knowledge deficit and negative attitudes regarding pain management [17].

Nonetheless, patients with SCD commonly report low self-esteem and feelings of hopelessness as a result of frequent pain, hospitalizations and loss of schooling (in children) and employment (in adults). These accounts could indicate depressive symptoms and should not be ignored by hematologists [12].

On the other hand, HRQOL may be overestimated by providers that do not regularly care for patients with SCD due to lack of understanding of the severity of the painful crises and the potential impact on function [19]. Studies reported that the more SCD pain a subject experienced, the worse the reported quality of life. One unit increase in pain (on a 0–9 scale) was associated with an approximate decrease of 1.4 (mental health) to 6 (both role functions) units on an SF-36 subscale [24].

Weakness

This study being cross sectional, does not allow inference of causality to be made. In addition, using convenient sample prevents generalizability of the study's results.

Sample size may not be large enough and could not be calculated due to unavailability of any official record of the number SCD sufferers in Bahrain. While finding an approval to collect a sample from SCD clinics went smoothly, the number of attendants in SMC and H.C clinics was an obstacle. Added to this, the self-report and recall biases were encountered.

Strengths

This research being the first study to address HRQoL in SCD in Bahrain, with a large enough sample size and the results offers a platform for further studies. Using two groups of respondents besides SCD allowed a valid comparison to be made which provided further strength of HRQOL in the SCD group in the society. Wide varieties of resources were used to collect a representative sample like the social network, health centers, and SMC. The questionnaire was valid in its English and Arabic versions with a high Cronbach's alpha in field studies. In this study the Cronbach's alpha was reliable in most HRQOL subscales (0.76- .89), except for Energy/Fatigue domains, Social Functioning and General Health which scored lower than 0.7. Appendix

Most importantly this study acknowledges a very important and sensitive issue in Bahrain, as the number of mortality associated with SCD attacks with sensational and evocative media reporting, which generates much resentment in the community against the Ministry of health. This research would help to understand the HRQOL in SCD and hence used to improve health services if needed. The results of the study can be used for further studies.

RECOMMENDATIONS

We would recommend conducting further researches to evaluate the factors that may influence the HRQoL in SCD patients like social support, coping skills, self-care ability, financial and housing support. In addition, a local study should be conducted in Bahrain to view sickle disease individual's knowledge and attitude toward treatment with hydroxyurea. Furthermore, there is an urgent need for a national SCD register to study and estimate the current prevalence of SCD in Bahrain.

ACKNOWLEDGMENT

The authors would like to acknowledge the grateful contribution of Dr. Mohamed Mandeel and the support from Dr. Ebtisam Fakhroo and family practice residency program administration. We also would like to thank Dr. Naeema Sabat and Dr. Waleed AlManea for their role in facilitating data collection from primary health centers and Salmaniya medical complex. We genuinely appreciate the efforts of the people who agreed to be part in the research.

APPENDIX

Table 1:

Step 1: Recording items

Item number	Change original response category[1]	To record value of:
1,2,20,22,34,36	1 →	100
	2 →	75
	3 →	50
	4 →	25
	5 →	0
3,4,5,6,7,8,9,10,11,12	1 →	0
	2 →	50
	3 →	100
13,14,15,16,17,18,19	1 →	0
	2 →	100
21,23,26,27,30	1 →	100
	2 →	80
	3 →	60
	4 →	40
	5 →	20
	6 →	0
24,25,28,29,31	1 →	0
	2 →	20
	3 →	40
	4 →	60
	5 →	80
	6 →	100
32,33,35	1 →	0
	2 →	25
	3 →	50
	4 →	75
	5 →	100

[1] Precoded response choices as printed in the questionnaire

Table 2:

Step 2: Averaging items to form scales

Scale	Number of items	After recoding per table 1 , average the following items :
Physical functioning	10	3 4 5 6 7 8 9 10 11 12
Role limitations due to physical health	4	13 14 15 16
Role limitation due to emotional problem	3	17 18 19
Energy / Fatigue	4	23 27 29 31
Emotional well-being	5	24 25 26 28 30
Social functioning	2	20 32
Pain	2	21 22
General Health	5	1 33 34 35 36

Table 3

Reliability, Central tendency and Variability of study scales

	Cronbach's Alpha	Mean	SD
Physical functioning	.889	67.29	26.04
Role-Physical	.828	54.54	40.45
Role-Emotional	.808	53.01	42.42
Energy_Fatigue	.670	50.62	19.50
Emotional Wellbeing	.752	61.02	19.59
Social Functioning	.640	68.42	24.16
Pain	.860	66.06	27.29
General Health	.489	57.82	19.01
Health change	-----	56.50	25.08

REFRENCES

1. Adekile, A. (2013). Sickle cell disease in Kuwait. Paper presented at the International Conference on Sickle Cell Disease Management and Prevention. 5-7; Manama: Kuwait.
2. Refaie, A. (2013). Pain management in SCD patients and the role of pain clinic. Paper presented at the International Conference on Sickle Cell Disease Management and Prevention. 5-7; Manama: Bahrain.
3. AlArrayed, S. (2013). Bahrain success story in controlling sickle cell disease 1984-2013. Paper presented at the International Conference on Sickle Cell Disease Management and Prevention. 5-7; Manama: Bahrain.
4. AlKhawaga, S. (2013). Predictors of risk of death in adult sickle cell patients admitted to intensive care unit in SMC. Paper presented at the International Conference on Sickle Cell Disease Management and Prevention. 5-7; Manama: Bahrain.
5. AlJufairi, Z. (2013). Maternal mortality among women with sickle cell disease in Kingdom of Bahrain between 1977 and 2012. Paper presented at the International Conference on Sickle Cell Disease Management and Prevention. Feb 5-7; Manama: Bahrain.
6. Mann-Jiles, V., & Morris, D. L. (2009). Quality of life of adult patients with sickle cell disease.J Am Acad Nurse Pract. 21(6), 340-349.
7. Jenerette, C. M., & Murdaugh, C. (2008). Testing the theory of self-care management for sickle cell disease. *Research in nursing & health*, *31*(4), 355-369.
8. Panepinto, J. A. (2012). Health-related quality of life in patients with hemoglobinopathies. *Hematology Am SocHematolEduc Program.* (1), 284-289.
9. Vilela, R. Q. B., Cavalcante, J. C., Cavalcante, B. F., Araújo, D. L., Lôbo, M. D. M., & Nunes, F. A. T. (2012). Quality of life of individuals with sickle cell disease followed at referral centers in Alagoas, Brazil. *Revista brasileira de hematologia e hemoterapia*, *34*(6), 442-446.
10. Amr, M. A. M., Amin, T. T., & Al-Omair, O. A. (2011). Health related quality of life among adolescents with sickle cell disease in Saudi Arabia. *Pan African Medical Journal*, *8*(1).
11. Dampier, C., LeBeau, P., Rhee, S., Lieff, S., Kesler, K., Ballas, S., ... & Wang, W. (2011). Health-related quality of life in adults with sickle cell disease (SCD): A report from the comprehensive sickle cell centers clinical trial consortium. *American journal of hematology*, *86*(2), 203-205.
12. Anie, K. (2005). Psychological complications in sickle cell disease. *Br J Haematol. 129*(6), 723-729.
13. McClish, D. K., Penberthy, L. T., Bovbjerg, V. E., Roberts, J. D., Aisiku, I. P., Levenson, J. L., ... &

Smith, W. R. (2005). Health related quality of life in sickle cell patients: the PiSCES project. *Health and Quality of Life Outcomes*, *3*(1), 1.
14. Jenerette, C. M. (2008). Relationships among types of social support and QOL in adults with sickle cell disease. *Southern Online Journal of Nursing Research*, *8*(3), 1-14.
15. Caird, H., Camic, P. M., & Thomas, V. (2011). The lives of adults over 30 living with sickle cell disorder. *British journal of health psychology*, *16*(3), 542-558.
16. Khader, S., Hourani, M. M., & Al-Akour, N. (2011). Normative data and psychometric properties of short form 36 health survey (SF-36, version 1.0) in the population of north Jordan/Données normatives et propriétés psychométriques du questionnaire d'évaluation de la santé SF-36 en version courte (SF-36, version 1.0) dans la population du nord de la Jordanie. *Eastern Mediterranean Health Journal*, *17*(5), 368.
17. Coons, S. J., Alabdulmohsin, S. A., Draugalis, J. R., & Hays, R. D. (1998). Reliability of an Arabic version of the RAND-36 Health Survey and its equivalence to the US-English version. *Medical care*, *36*(3), 428-432.
18. RAND Health [Internet]. Medical Outcomes Study: 36-Item Short Form Survey Scoring Instructions. California: RAND; [cited 2013 Oct 25] available from: http://m.rand.org/health/surveys_tools/mos/mos_core_36item_scoring.html
19. Thomas, V. J., & Taylor, L. M. (2002). The psychosocial experience of people with sickle cell disease and its impact on quality of life: Qualitative findings from focus groups. *British journal of health psychology*, *7*(3), 345-363.
20. Anie, K. A., Grocott, H., White, L., Dzingina, M., Rogers, G., & Cho, G. (2012). Patient self-assessment of hospital pain, mood and health-related quality of life in adults with sickle cell disease. *BMJ open*, *2*(4), e001274.
21. Darwish, A. (2013). Psycho-social problems vs. narcotic drug dependency in sickle cell pts. With frequent visits to accidents & emergency. Paper presented at the 1st International Conference on Sickle Cell Disease Management and Prevention, 5-7; Manama: Bahrain.
22. Steinberg, M. H., Barton, F., Castro, O., Pegelow, C. H., Ballas, S. K., Kutlar, A., ... & Varma, M. (2003). Effect of hydroxyurea on mortality and morbidity in adult sickle cell anemia: risks and benefits up to 9 years of treatment. *Jama*, *289*(13), 1645-1651.
23. Ballas, S. K., Bauserman, R. L., McCarthy, W. F., Castro, O. L., Smith, W. R., Waclawiw, M. A., & Investigators of the Multicenter Study of Hydroxyurea in Sickle Cell Anemia. (2010). Hydroxyurea and acute painful crises in sickle cell anemia: effects on hospital length of stay and opioid utilization during hospitalization, outpatient acute care contacts, and at home. *Journal of pain and symptom management*, *40*(6), 870-882.
24. Ballas, S. K., Barton, F. B., Waclawiw, M. A., Swerdlow, P., Eckman, J. R., Pegelow, C. H., ... & Bonds, D. R. (2006). Hydroxyurea and sickle cell anemia: effect on quality of life. *Health and Quality of Life Outcomes*, *4*(1), 1.
25. Asnani, M. R., Reid, M. E., Ali, S. B., Lipps, G., & Williams-Green, P. (2008). Quality of life in patients with sickle cell disease in Jamaica: rural-urban differences. *Rural Remote Health*, *8*(2), 890.
26. Ohaeri, J. U., & Shokunbi, W. A. (2002). Psychosocial burden of sickle cell disease on caregivers in a Nigerian setting. *Journal of the National Medical Association*, *94*(12), 1058.
27. Ratanawongsa, N., Haywood, C., Bediako, S. M., Lattimer, L., Lanzkron, S., Hill, P. M., ... & Beach, M. C. (2009). Health care provider attitudes toward patients with acute vaso-occlusive crisis due to sickle cell disease: Development of a scale. *Patient education and counseling*, *76*(2), 272-278.
28. Glassberg, J. A., Tanabe, P., Chow, A., Harper, K., Haywood, C., DeBaun, M. R., & Richardson, L. D. (2013). Emergency provider analgesic practices and attitudes toward patients with sickle cell disease. *Annals of emergency medicine*, *62*(4), 293-302.