SUBMITTED 14 DEC 23
REVISION REQ. 17 JAN 24; REVISION RECD. 30 JAN 24
ACCEPTED 18 FEB 24
ONLINE-FIRST: MARCH 2024
DOI: https://doi.org/10.18295/squmj.3.2024.019

Health-Related Quality of Life of Omani Adult Patients with Sickle Cell Disease at the Sultan Qaboos University Hospital

Sara Al-Hinai,[1] Asma Al-Rashdi,[1] *Hana Al-Sumri[2]

[2]Department of Family Medicine and Public Health, [1]College of Medicine and Health Sciences, Sultan Qaboos University, Muscat, Oman.
**Corresponding Author's e-mail: alsumry@squ.edu.om*

Abstract

Objectives: The quality of life of Omani adults with sickle cell disease (SCD) is not yet known. This study aimed to determine the health-related quality of life (HRQoL) of adult Omani patients with SCD attending a single tertiary center. ***Methods:*** A cross-sectional study was conducted among patients with SCD from June to October 2022 at the Sultan Qaboos University Hospital. A validated Arabic version of the 36-Item Short-Form Health Survey (SF-36) was used to assess HRQoL over eight domains. ***Results:*** A total of 235 patients with SCD participated in the study, of which the majority were female (74.9%) and 18–35 years of age (64.6%). The lowest HRQoL was reported for the domain of role limitations due to physical health (median score: 25.0; interquartile range [IQR]: 35.0), while the highest was seen for role limitations due to emotional problems (median score: 66.7, IQR: 100.0). Frequent SCD-related emergency department visits/hospital admissions and the adverse effect of SCD on patients' social lives had a significant negative impact on SF-36 scores for all eight HRQoL domains ($p \leq 0.05$). In addition, the impact of SCD on academic performance and a history of having been bullied due to SCD had a significant negative impact on SF-36 scores for several domains ($p \leq 0.05$). **Conclusion:** Omani adult patients with SCD reported relatively poor HRQoL in several domains, with various factors found to have significant associations with poorer HRQoL. Healthcare providers and policy-makers

should consider offering additional clinical, educational, and financial support to patients to enhance their HRQoL.

Keywords: Sickle Cell Disease; Health-Related Quality of Life; Adults; Oman.

Advances in Knowledge:

- Quality of life of Omani patient with SCD was not known and has been measured in this study using a validated Arabic version of the widely used SF-36 quality of life survey.
- Study findings indicate a poor quality of life among adult patients with SCD in Oman in several domain of the SF-36 survey
- Frequent SCD-related emergency department admissions, the adverse effect of SCD on patients' social lives, the impact of SCD on academic performance and a history of having been bullied due to SCD had a significant negative impact on SF-36 scores for several domains.

Application to Patient Care:

- Healthcare providers should include early screening for SCD-related complications, to reduce the need for frequent hospitalization.
- Policy-makers should consider introducing protective and supportive legislation to reduce social stigma surrounding this disease thus improving the social life of patients, their academic performance and encourage acceptance of this patient group within the workforce and community.

Introduction

Hemoglobinopathies represent a significant public health issue worldwide; this group of inherited blood disorders causes abnormalities in the synthesis and structure of hemoglobin molecules, reducing the oxygen-carrying capacity of the red blood cells.[1,2] Among these, sickle cell disease (SCD) is a very prevalent monogenic hemoglobinopathy which occurs due to the substitution mutation of thiamine for adenine in the beta-chain gene, leading to the creation of hemoglobin S, a variant of normal hemoglobin.[3,4] This mutation causes hemoglobin polymerization, resulting in irregular, crescent-shaped, sticky, rigid, and fragile red blood cells.[4] Patients experience a wide range of complications, including chronic pain, fatigue, cardiovascular events, leg ulcers, and recurrent, unpredictable episodes of microcirculation obstruction resulting in multi-organ ischemic damage and excruciating

sickle cell-related pain, also referred to as vaso-occlusive crises.[5] Other possible complications include stroke, pulmonary hypertension, acute chest syndrome, gallstones, priapism, and pregnancy complications.[5,6]

Due to the chronic nature of SCD and severity of SCD-related complications, affected individuals experience considerable disruption of their normal day-to-day lives and require frequent medical attention in the form of emergency department (ED) visits or hospitalization.[7,8] This burden is often compounded by a lack of awareness of disease complications and underestimation of patients' needs by medical professionals, sometimes resulting in an over-reliance on ED care.[9] Such factors contribute to poor health-related quality of life (HRQoL), a concept defined as the "patient's appraisal of how his/her well-being and level of functioning, compared to the perceived ideal, are affected by individual health".[10] Research shows that adult patients with SCD have poorer HRQoL compared to the general population as well as individuals with other chronic diseases.[12–13] In addition, researchers have observed that SCD adversely impacts various aspects of patients' lives, causing sleep disturbances and an inability to maintain their job and academic performance as well as partake in normal social, recreational, and daily activities, thereby affecting their marital, financial, and employment prospects.[8,13]

A study among 629 Saudi adult patient with SCD reported patients with disease related complications such as; fever, skin redness, swelling, or history of blood transfusion tended to adversely impact the health related quality of life using the SF-36 scale.[12] In Oman, hemoglobinopathies are relatively common; the prevalence rate of SCD is reported to be 0.2%, while approximately 4.8–6.0% of the population are carriers of the sickle cell trait.[14] A recent study to assess health-related stigma, social support, self-care, and self-efficacy among Omani adults with SCD found that most participants experienced a low level of stigma due to the social and psychological support provided by their families and their strong Islamic faith, although unmarried participants reported greater stigma in contrast to married participants.[15] Nonetheless, there remains limited data concerning the HRQoL of Omani adults with hemoglobinopathies. Such research is crucial to reduce the burden on health resources and to create policies to enhance the lives of affected patients and aid healthcare professionals in counselling and managing this chronic disease. This study therefore aimed to assess the HRQoL of Omani adults with SCD using The Medical Outcomes 36-item Short-Form Health Survey (SF-36). The study will determine the related associations of patient's measured

HRQoL with selected demographic (including; age, gender, education level and employment status) social (including; impact on marriage, perceived disease impact on social and academic life) and clinical characteristics (including; frequency of hospital admissions, blood transfusion, disease related complications).[12]

Methods

Study Design and Setting

A cross-sectional study was conducted from June to October 2022 at the Sultan Qaboos University Hospital (SQUH), a single tertiary care center in Muscat, Oman. The target population of the study consisted of adult patients of both genders and of Omani nationality aged 18 years or older who had been diagnosed with SCD and who were being followed-up at SQUH over the course of the study period. Patients with other diagnosed haemoglobinopathies were excluded from the study.

Sample Size Calculation

Based on an estimated total number of patients with SCD being followed-up at SQUH (N = 1,000 registered patients in 2022), the necessary sample size for this study was calculated to be 215, taking into account a confidence interval of 95% and a permissible rate of error of 5%. The mean and standard deviation (SD) of the outcome measure was taken from a previous study conducted in a neighboring country.[13]

Study instrument

The Medical Outcomes 36-item Short-Form Health Survey (SF-36) is a standardized, validated questionnaire to assess HRQoL over eight domains with a total of 36 items , including the respondent's perceptions of their physical function (10 items); physical role health (4 items); emotional role functions (3 items); emotional wellbeing (5 items); social function (2 items); bodily pain (2 items);energy/fatigue (5 items) and general health perceptions (5 items).[16]

Data from SF-36 were scored based on the scoring system reported by RAND Health. Each item in the SF-36 questionnaire is represented as a single variable and scaled from 0 to 100, with a higher score indicative of better HRQoL.[21] The original SF-36 has been shown to be a reliable measure of HRQoL, with Cronbach's alpha values ranging from 0.78–0.93[17,18,19] . The internal consistency and reliability of the SF-36 has been investigated in a previous pilot

study of 80 patients with SCD in Saudi Arabia. High internal consistency (Cronbach's alpha > 0.6) have been reported for physical function (Cronbach's alpha = 0.81); physical role health (Cronbach's alpha = 0.84); emotional role functions (Cronbach's alpha = 0.86); vitality (Cronbach's alpha = 0.79); emotional wellbeing (Cronbach's alpha = 0.67); social function (Cronbach's alpha = 0.67); bodily pain (Cronbach's alpha = 0.84); and general health (Cronbach's alpha = 0.60)[12]. For the purposes of the present study, this former validated Arabic-language version of the SF-36 was used.[12]

Another two-part questionnaire was developed to collect data regarding the participants' demographic, social, and clinical characteristics. The first section assessed the demographic background of the participants, including their gender, age, place of residence, education , employment and marital status. The second section collected information regarding the participants' clinical characteristics and the perceived social impact of the disease. This included a recent history of visits to the ED or hospital admissions over the past three months, presence of medical complications, existence of family support, and the perceived impact of the disease on their academic/work performance, among other items.

Data Collection Process

A link to the online survey distributed electronically to all eligible patients with SCD identified using the hospital registry list. The questionnaire's QR code was shared with participants and it was self-administered. All Omani adult patients with diagnosed SCD and were following up in the hematology department outpatient clinic during the study period were invited into the study. A study information sheet was provided for participants in the preferred language prior to questionnaire distribution. In addition a consent statement attached to the questionnaire link had to be ticked by participants who have agreed to take part in the study. The exclusion criteria included: patients with SCD <18 years old , patients who cannot write or read, patients admitted due to vaso-oclusive crisis or surgery or those who were under sedative painkillers. To avoid duplicated data, a researcher ensured that each participant filled out the questionnaire only once using each participant's date of birth as an identifier. Ethical approval for this study was obtained from the Medical Research and Ethics Committee (MREC) of the College of Medicine and Health Sciences, Sultan Qaboos University.

Data Analysis and Interpretation

Descriptive data were presented as percentages for categorical variables. Continuous variables were presented as either means and SDs or medians and interquartile ranges, depending on the normality of the data distribution as determined using a one-sample Kolmogorov-Smirnov test. Scores for each of the eight HRQoL domains were calculated using the SF-36 scoring index and presented as either means or medians, depending on the normality of the data distribution. Bivariate associations between mean/median SF-36 scores for each of the HRQoL domains and various demographic, clinical, and social impact factors were calculated using either a parametric or a non-parametric statistical tests. Analyses were conducted using the Statistical Package for the Social Sciences (SPSS) (IBM Corp., Armonk, NY). A p value of ≤ 0.05 was considered statistically significant.

Results

Sociodemographic and Clinical Characteristics

A total of 235 Omani adult patients with SCD participated in the study, of which the majority were female (74.9%) and aged 18–25 (26.5%) or 25–35 years old (38.1%). Most patients resided in either Muscat (39.7%) or Ad Dhakhiliyah Governorate (23.7%). The majority were married (53.4%), with only two patients being divorced (0.8%) and one (0.4%) widowed; the remaining 45.3% were single. In terms of education, 69.1% of patients had a college diploma, bachelor's degree, or postgraduate qualification. Over one-third of the sample were unemployed (14.6%) or currently seeing work (24.9%).

In terms of clinical characteristics, most patients (61.3%) had a history of recent ED visits or hospital admissions due to SCD, with 35.8%, 13.2%, 3.3%, and 9.0% reporting 1–2, 3–4, 5–6, and ≥7 visits/admissions in the preceding three months, respectively. Additionally, 46.5% of patients had received at least one exchange transfusion, while 16.2% had undergone a splenectomy [Table 1]. Fatigue was the most frequently reported SCD-related disease complication (80.5%), followed by bone/joint problems (41.0%), gallstones (32.4%), recurrent infections (27.1%), splenomegaly (26.2%), hip/shoulder necrosis (22.9%), and acute chest syndrome (21.9%) [Figure 1]. In addition, the majority of married female patients (68.7%) reported having had difficulties conceiving or complications during pregnancy. A proportion of patients also reported mental health difficulties, with anxiety being most common (23.0%), followed by sleep disturbances (18.7%), depression (11.9%), and social isolation (6.8%).

Social Impact of the Disease

With regards to the social impact of SCD, 44.6% of single respondents reported that the disease had played some role in delaying marriage, although only 24.8% of married participants claimed that SCD had caused them marital issues. Just over one-third (36.8%) of patients with children stated that SCD had negatively affected their relationship with their children or their ability to care for them. Most patients reported that SCD had negatively affected their academic performance (64.9%) and limited their social life (64.2%). In addition, 34.5% claimed that they had been bullied or had their abilities belittled due to SCD [Table 2].

Health-Related Quality of Life Domains

Of the eight HRQoL domains assessed in the SF-36 tool, the highest median score (66.7) was reported for the domain of role limitations due to emotional problems, indicating that the patients experienced better quality of life in this domain. The next best HRQoL domain was social functioning (median SF-36 score: 62.5), followed by physical functioning and emotional well-being (median SF-36 scores: 60.0 each). In turn, the domain with the lowest median score, denoting poorest HRQoL, was role limitations due to physical health (median SF-36 score: 25.0), followed by energy/fatigue (median SF-36 score: 50.0). Patients reported relatively neutral/better quality of life for the remaining two HRQoL domains, bodily pain and general health (median SF-36 scores: 55.0 each) [Table 3].

Bivariate Analysis

Overall, no significant associations were observed between any of the selected sociodemographic characteristics and the eight individual HRQoL domains, apart from employment status in which a significant association was observed with the social functioning domain ($p = 0.009$) [Table 4]. However significant associations were seen between number of disease complications and six HRQoL domains, with patients with a greater number of complications reporting poorer HRQoL in domains of physical functioning ($p = 0.005$), bodily pain ($p = 0.003$), general health ($p = 0.026$), social functioning ($p = 0.006$), and role limitations due to physical health ($p = 0.014$) and emotional problems ($p = 0.018$).

A history of pregnancy difficulties was significantly associated with lower scores for the HRQoL domains of pain (p <0.001), general health (p = 0.007), and role limitations due to physical health (p = 0.032). Patients with a history of exchange transfusions demonstrated significantly poorer scores for the physical functioning (p = 0.020), energy/fatigue (p = 0.033), pain (p <0.001), general health (p = 0.043), social functioning (p = 0.046), and role limitations due to physical health (p = 0.019) domains, while those with a history of blood transfusions reported worse bodily pain (p = 0.007). Number of recent ED visits/ hospital admissions was a crucial factor found to be associated with significantly poorer HRQoL across all eight domains ($p \leq 0.05$ each).

Interestingly, however, patients who reported being compliant with their prescribed medications (p = 0.021) and those who received family support (p = 0.006) reported poorer scores for the pain HRQoL domain. Patients who exercised more frequently reported better HRQoL for the physical functioning domain (p = 0.031). In terms of the perceived social impact of the disease, both patients who reported adverse academic performance and those who had experienced bullying due to SCD reported significantly poorer scores in all HRQoL domains apart from physical functioning (p <0.05 each). Importantly, the perception that SCD had resulted in limitations to the patients' social lives was significantly associated with all eight HRQoL domains ($p \leq 0.05$) [Table 5].

Discussion

Research focusing on HRQoL is essential to ensure the provision of personalized and effective patient care, reduce the onset of complications, advocate for policy changes, and ultimately help improve the lives of individuals living with chronic conditions, including SCD.[20,21] As such, this study aimed to describe the HRQoL of Omani adult patients with SCD and determine associations with various sociodemographic and clinical characteristics.

Of all eight assessed HRQoL domains, the present study revealed that Omani adults with SCD reported the poorest quality of life in the domain of role limitations due to physical health (median SF-36 score: 25.0). A similar study reported comparable findings, in which this domain received the lowest score (mean SF-36 score: 35.9) among adult patients with SCD attending two hospitals in two different regions of Saudi Arabia.[12] On the other hand, the studies differenced as Omani patients with SCD in the current study reported the highest quality of life in the domain of role limitations due to emotional problems (median SF-36

score: 66.7), while the Saudi Arabian study reported the best HRQoL for the domain of social functioning (mean SF-36 score: 60.1), the second highest domain in the present study (median SF-36 score: 62.5).[12] In general, however, Omani patients in our study reported generally poor HRQoL in multiple domains, considering a cut-off score of 50 to represent a distinction point between poorer and better HRQoL.[16,22]

As with most Arab countries, the practice of early and universal marriage is still highly prevalent in Oman; essentially, this means that the vast majority of individuals aged >20 years are expected to be married.[23] Thus, the fact that almost half of the sample (45.3%) in the present study were single represents a considerable deviation of sociocultural norms. The study from Saudi Arabia similarly reported that 55.6% of adult patients with SCD were unmarried, although another study indicated a much lower rate of single participants (27.5%) among a cohort of patients with SCD attending multiple primary care centers in Bahrain.[12,24] Nonetheless, despite the high prevalence of unmarried patients, marital status was not found to be significantly associated with HRQoL in the current study. Similarly, no associations were determined with most other sociodemographic factors, such as gender and age; this is consistent with the findings of the aforementioned Saudi Arabian study.[12] Previous research has indicated that age might be related to poorer quality of life in adolescents with SCD, particularly in domains related to social functioning, possibly due to higher rates of actual or perceived stigmatization by their peers.[25,27] In contrast, another study noted that, among a cohort of young adults with SCD, increasing disease severity trajectories over time were associated with older age, a factor linked with higher risk of SCD complications and increased care requirements, likely resulting in poor HRQoL.[28]

Indeed, frequent complications of SCD seen in the current study included fatigue (80.5%), bone/joint problems (40.1%), and gallstones (32.4%), findings relatively similar to a previous study conducted in the USA which reported frequent digestive and musculoskeletal complications among patients with SCD.[29] As expected, a greater number of complications, more frequent ED visits or hospitalizations, and a history of blood/exchange transfusions were factors found to significantly worsen HRQoL in the present study, all of which can be considered indications of disease severity. In addition, the present study found that the majority (68.7%) of married, female patients with SCD complained of difficulties conceiving and/or pregnancy complications, with significantly lower quality of life in several domains, including general health, pain, and role limitations due to physical function.

Moreover, the present study identified a significant association between employment status and scores for the HRQoL domain of social functioning, in which unemployed patients reported significantly poorer scores compared to those who were employed, students, or seeking a job. This finding is supported by previous research indicating that unemployment is significantly linked to worse social functioning among patients with SCD.[28,29] It is believed that employment is a crucial mediator of quality of life among patients with chronic diseases, likely because it impacts multiple domains of life, including providing individuals with a sense of purpose and value, fostering personal identity and self-esteem, and promoting financial independence and material security.[30] Despite frequent hospital admissions and complications, SCD did not appear to have affected the current study's patients' level of academic attainment, given that the majority (69.8%) had achieved a college diploma or higher degree; however, the high rate of unemployment or job-seeking (39.5%) indicates that securing a job remained a challenge because of their medical condition. Moreover, although no direct association was observed between HRQoL and education level *per se*, patients reported a significant impact with regards to the perceived negative impact of the disease on academic performance, including missing classes, inability to participate in various school activities, and performing poorly academically due to their medical condition.

Overall, patients who reported social restrictions due to SCD also reported significantly poorer HRQoL across all eight domains. This is likely as a result of overlap and interactions between various HRQoL domains; for instance, a previous study reported a significant association between pain and social functioning.[29] This is understandable as patients in greater pain, indicating a more severe disease course, are likely to have lower physical functioning, vitality, general health, and emotional well-being as a result, all of which can impact the patients' social lives in various ways. Al-Raqaishi *et al.* noted that patients with SCD often suffered the effects of health-related stigma.[15] Similarly, just over one-third (34.5%) of patients in the present study reported having been bullied or belittled due to their condition, a factor which significantly affected various HRQoL domains. This is comparable to another study conducted in Nigeria, in which 26% of adults with SCD experienced teasing or bullying, with 66% perceiving negative societal attitudes towards the disease.[26]

To the best of the authors' knowledge, this study is the first in Oman to measure the HRQoL of Omani adult patients with SCD. However, certain limitations should be acknowledged.

The data collection tool was self-administered, and therefore dishonest responses cannot be discounted. Additionally, the study was conducted in a single center; thus, the results might not be representative of all Omani adult patients with SCD.

Conclusion

The studied sample of Omani adult patients with SCD reported generally poor quality of life in multiple HRQoL domains, particularly in the domains of role limitations due to physical health and energy/fatigue. In addition, significant associations were observed between poorer HRQoL and various clinical and social characteristics. Such findings indicate that healthcare providers should devote more attention to patients with SCD during consultations, including screening early for complications, to enhance patients HRQoL. In addition, policy-makers should consider introducing protective legislation to reduce social stigma surrounding this disease and encourage acceptance of this patient group within the workforce and community.

Authors' Contribution

HS conceptualized the study and supervised the work. SH and AR collected the data. All authors analysed the data and drafted the manuscript. All authors approved the final version of the manuscript.

Acknowledgement

The authors would like to extend their gratitude to the department of Hematology at Sultan Qaboos University Hospital for facilitating data collection and to patients with sickle cell disease for their participation in the study.

Conflicts of Interest

The authors declare no conflict of interests.

Funding

No funding was received for this study.

References

1. Williams TN, and Weatherall DJ. World distribution, population genetics, and health burden of the hemoglobinopathies. Cold Spring Harb Perspect Med 2012; **2**(9), p. a011692. doi: 10.1101/cshperspect.a011692.

2. Indrák K, Divoká M, Pospíšilová D, Čermák J, Beličková M, Horváthová M, et al. [Hemoglobinopathies]. Vnitr Lek 2018; **64**(5):476–487.
3. Kavanagh P L, Fasipe TA, and Wun T. Sickle cell disease: a review. *JAMA* 2022; **328**(1):57–68. doi: 10.1001/jama.2022.10233.
4. Sundd P, Gladwin MT, and Novelli EM. Pathophysiology of sickle cell disease. *Annual Review of Pathology. WIREs Mech Dis* 2019; **14**(2019): 263–292. doi: 10.1146/annurev-pathmechdis-012418-012838.
5. Ware RE, de Montalembert M, Tshilolo L, and Abboud MR. Sickle cell disease. *The Lancet* 2017; **390**(10091): 311–323. doi: 10.1016/S0140-6736(17)30193-9
6. Ogu UO, Badamosi NU, Camacho PE, Freire AX, and Adams-Graves P. Management of sickle cell disease complications beyond acute chest syndrome. J Blood Med, 2021; **12**: 101–114. doi: 10.2147/JBM.S291394.
7. de O da P, Menezes AS, Len CA, Hilário MOE, Terreri MTRA, and Braga JAP. Quality of life in patients with sickle cell disease. Rev Paul Pediatr 2013; **31**: 24–29. doi: 10.2147/JPR.S261605.
8. Rizio AA, Bhor M, Lin X, McCausland KL, White MK, PauloseJ, et al. The relationship between frequency and severity of vaso-occlusive crises and health-related quality of life and work productivity in adults with sickle cell disease. Qual Life Res 2020; **29**(6):1533–1547. doi: 10.1007/s11136-019-02412-5.
9. Smeltzer MP, Howell KE, Treadwell M, Preiss L, King A., Glassberg JA, et al. Identifying barriers to evidence-based care for sickle cell disease: results from the Sickle Cell Disease Implementation Consortium cross-sectional survey of healthcare providers in the USA. *BMJ Open* 2021; 11, p. e050880. doi: 10.1136/bmjopen-2021-050880.
10. Panepinto J., and Bonner, M. Health-related quality of life in sickle cell disease: past, present, and future. Pediatr Blood Cancer 2012; **59**(2):377–385. doi: 10.1002/pbc.24176.
11. Bhagat VM, Baviskar SR, Mudey AB, and Goyal RC. Poor health related quality of life among patients of sickle cell disease. Indian J Palliat Care 2014; **20**(2): 107–111. doi: 10.4103/0973-1075.132622.
12. Ahmed AE, Alaskar AS, Al-Suliman A, Jazieh AR, McClish DK, Al Salamah M, et al. Health-related quality of life in patients with sickle cell disease in Saudi Arabia. Health Qual Life Outcomes 2015; **13**(1):183. doi: 10.1186/s12955-015-0380-8.

13. Khaled A,AL Mghaslah D,Mutiq R, Al Shehri W. Sickle cell disease patients' health-related quality of life in the southern region of Saudi Arabia. Int J Clin Pract 2021; **75**(4), p. e13775. doi: 10.1111/ijcp.13775.
14. Al Riyami AA, Suleiman AJ, Afifi M, Al-Lamki ZM, and Daar S. A community-based study of common hereditary blood disorders in Oman. East Mediterr Health J 2001; **7**(6) :1004–1011.
15. Al Raqaishi H, Al Qadire M, Alzaabi O, and Al Omari O. Health-related stigma, social support, self-efficacy, and self-care actions among adults with sickle cell disease in Oman. Clin Nurs Res 2022; **31**(5) :803–811. doi: 10.1177/10547738211046731.
16. Ware JE Jr, and Sherbourne CD .The MOS 36-ltem short-form health survey (SF-36): I. Conceptual framework and item selection. *Medical Care* 1992; **30**(6):473–483.
17. McHorney CA, Ware JA, and Raczek AE. The MOS 36-item short-form health survey (SF36): II. Psychometric and clinical tests of validity in measuring physical and mental health constructs. *Medical Care* 1993; **31**(3) :247–263. doi: 10.1097/00005650-199303000-00006.
18. Garratt AM, Ruta DA, Abdalla MI, Buckingham JK, and Russell IT. The SF36 health survey questionnaire: an outcome measure suitable for routine use within the NHS. BMJ Med 1993; **306**(6890) :1440–1444. doi: 10.1136/bmj.306.6890.1440.
19. McClish DK, Penberthy LT, Bovbjerg VE, Roberts JD, Aisiku IP, Levenson JL, et al. Health related quality of life in sickle cell patients: the PiSCES project. Health Qual Life Outcomes 2005; **3** : 50. doi: 10.1186/1477-7525-3-50.
20. de Wit M, and Hajos T. Health-related quality of life. In: Gellman, M.D., and Turner, J.R. (Editors). *Encyclopedia of Behavioral Medicine* 2013; Springer, New York, NY. P 929–931. doi: 10.1007/978-1-4419-1005-9_753.
21. Treadwell MJ, and Anie KA. Quality of life in sickle cell disease: what matters. Hematol Oncol Clin North Am 2022; **36**(6) :1137–1149. doi: 10.1016/j.hoc.2022.06.010.
22. Wagner AK, Gandek B, Aaronson NK, Acquadro C, Alonso J, Apolone G, et al. Cross-cultural comparisons of the content of SF-36 translations across 10 countries: results from the IQOLA project. J Clin Epidemiol 1998; **51**(11) :925–932. doi: 10.1016/s0895-4356(98)00083-3 10.1016/s0895-4356(98)00083-3.

23. Islam MM, Dorvolo AS, and Al-Qasmi AM. The pattern of female nuptiality in Oman. Sultan Qaboos Univ Med J 2013; **13**(1) :32–42. doi: 10.12816/0003193.
24. AlSaleh A, Elbarbary A, Al Qashar A, Alserdieh F, Alahmed F, Alhaddar H, et al. Determinants of the quality of life of patients with sickle cell disease in Bahrain: implications for a patient-centered management approach at the primary health care. Bahrain Med Bull 2021; **43**(2) : 431–437.
25. Adeyemo TA, Ojewunmi OO, Diaku-Akinwumi IN, Ayinde OC, and Akanmu AS. Health related quality of life and perception of stigmatisation in adolescents living with sickle cell disease in Nigeria: a cross sectional study. Pediatr Blood Cancer 2015; **62**(7) : 1245–1251. doi: 10.1002/pbc.25503.
26. Anie KA, Egunjobi FE, and Akinyanju OO. Psychosocial impact of sickle cell disorder: perspectives from a Nigerian setting. Global Health 2010; **6**, p. 2. doi: 10.1186/1744-8603-6-2.
27. Kayle M, Docherty SL, Sloane R, Tanabe P, Maslow G, Pan W, et al. Transition to adult care in sickle cell disease: a longitudinal study of clinical characteristics and disease severity. Pediatr Blood Cancer 2019; **66**(1), p. e27463. doi: 10.1002/pbc.27463.
28. Knisely MR, Pugh N, Kroner B, Masese R, Gordeuk V, King AA, et al. Patient-reported outcomes in sickle cell disease and association with clinical and psychosocial factors: report from the Sickle Cell Disease Implementation Consortium. Am J Hematol 2020; **95**(9) : 1066–1074. doi: 10.1002/ajh.25880.
29. Treadwell MJ, Mushiana S, Badawy SM, Preiss L, King AA, and Kroner B, et al. An evaluation of patient-reported outcomes in sickle cell disease within a conceptual model. Qual Life Res 2022; **31**(9) :2681–2694. doi: 10.1007/s11136-022-03132-z.
30. Andreu Y, Picazo C, Murgui S, Soto-Rubio A, García-Conde A, and Romero R. Exploring the independent association of employment status to cancer survivors' health-related quality of life. Health Qual Life Outcomes 2023; **21**(1), p. 44. doi: 10.1186/s12955-023-02124-y.

Table 1: Demographic and clinical characteristics of Omani adult patients with sickle cell disease (N = 235).

Characteristic	n (%)*
Gender	
Male	59 (25.1)
Female	176 (74.9)
Age (years)	
18–25	60 (26.5)
25–35	86 (38.1)
35–45	54 (23.9)
>45	26 (11.5)
Place of residence	
Muscat	92 (39.7)
Ad Dhakhiliyah	55 (23.7)
Al Batinah	39 (16.8)
Ash Sharqiyah	30 (12.9)
Ad Dhahirah	10 (4.3)
Al Buraimi	4 (1.7)
Dhofar	2 (0.9)
Marital status	
Single	107 (45.3)
Married	125 (53.4)
Divorced	2 (0.8)
Widowed	1 (0.4)
Education level	
Elementary	3 (1.3)
Middle school	7 (3.0)
High school	61 (26.0)
Diploma	45 (19.1)
Bachelor	103 (43.8)
Postgraduate	16 (6.8)
Employment status	
Student	41 (17.6)
Self-employed	9 (3.9)
Employed	77 (33.0)
Unemployed	
Seeking a job	
Retired	14 (6.0)
Recent† ED visits/hospital admissions due to SCD	
0	82 (38.7)
1–2	76 (35.8)
3–4	28 (13.2)
5–6	7 (3.3)
≥7	19 (9.0)
Hx of exchange transfusions	
Yes	107 (46.5)
No	123 (53.5)
No. of exchange transfusions	
1–2	44 (42.7)
3–4	21 (20.4)

≥5	38 (36.9)
Hx of splenectomy	
Yes	37 (16.2)
No	191 (83.8)

ED = emergency department; SCD = sickle cell disease; Hx = history.

**Percentages were calculated from the total number of valid responses due to missing data for certain variables (<5%). †Within the past 3 months.*

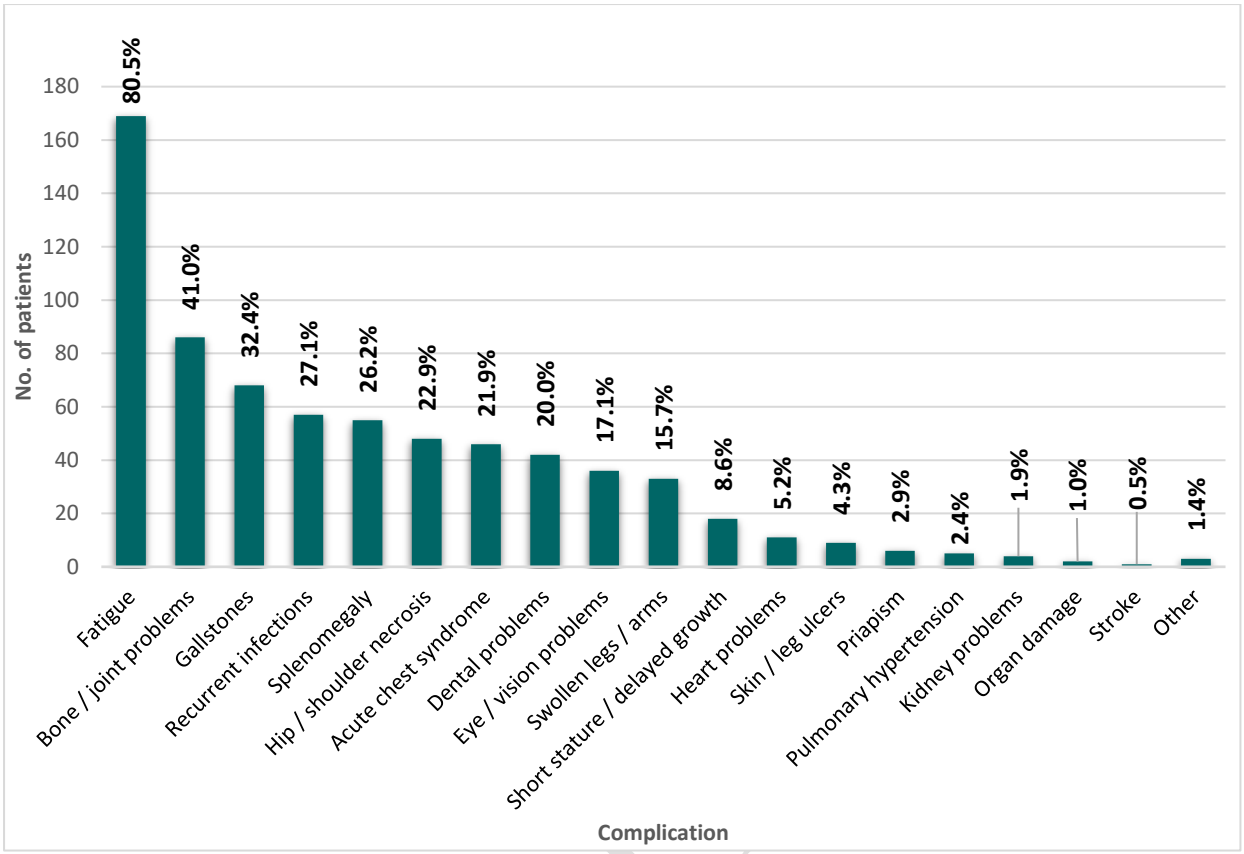

Figure 1: Frequency of sickle cell disease-related complications among Omani adult patients with sickle cell disease (N = 235).

Note: Percentages do not add up to 100% as patients may have had more than one complication.

Table 2: Frequency of responses to survey items relating to the social impact of sickle cell disease among Omani adult patients with sickle cell disease (N = 235).

Item	n (%)*
If not yet married, was SCD a reason for delaying marriage?	
Yes	45 (44.6)
No	56 (55.4)
If married, has SCD caused any marital issues?	
Yes	27 (24.8)
No	82 (75.2)
If you have children, has SCD negatively affected your relationship or your ability to take care of them?	
Yes	32 (36.8)
No	55 (63.2)
Has SCD impacted your academic performance (e.g., absences/lower grades/failing/not participating in school activities)?	
Yes	146 (64.9)
No	79 (35.1)
Has SCD limited your social life (e.g., attending gatherings/parties with family and friends/travelling/participating in sports/clubs/other hobbies)?	
Yes	145 (64.2)
No	81 (35.8)
Have you been bullied/belittled by others due to SCD?	
Yes	78 (34.5)
No	148 (65.5)

SCD = sickle cell disease.

**Percentages were calculated from the total number of valid responses due to missing data for certain variables (<5%).*

Table 3: Health-related quality of life scores* per domain among Omani adult patients with sickle cell disease (N = 235).

HRQoL domain	Score	
	Median†	IQR
Physical functioning	60.0	35.0
Role limitations due to physical health	25.0	75.0
Role limitations due to emotional problems	66.7	100.0
Energy/fatigue	50.0	25.0
Emotional wellbeing	60.0	27.0
Social functioning	62.5	38.0
Pain	55.0	43.0
General health	55.0	25.0

IQR = interquartile range.

**Assessed using the 36-Item Short-Form Health Survey. †Scores were presented as medians as the data did not follow normal distribution according to a one-sample Kolmogorov-Smirnov test.*

493 **Table 4:** Associations between selected sociodemographic variables and health-related quality of life domain scores* among Omani
494 adult patients with sickle cell disease (N = 235).

Variable		HRQoL domain, Mean score ± SD† or mean rank							
		Physical functioning	**Energy/fatigue**	**Pain**	**General health**	**Role limitations due to physical health**	**Role limitations due to emotional problems**	**Emotional wellbeing**	**Social functioning**
Gender	Male	114.41	100.97	103.71	95.11	100.11	112.59	105.90	101.16
	Female	101.88	105.74	110.84	113.07	105.31	107.14	108.70	107.62
	p value	0.194‡	0.615‡	0.462‡	0.064‡	0.574‡	0.557‡	0.774‡	0.496‡
Age (years)	18–25	106.67	105.10	57.98 ± 28.3	51.47 ± 16.9	106.41	106.47	101.91	110.76
	25–35	98.36	96.55	57.56 ± 26.8	55.06 ± 16.4	101.38	106.41	104.81	104.12
	35–45	99.59	99.04	51.01 ± 27.9	52.98 ± 18.2	96.91	91.76	100.17	86.71
	>45	95.08	108.58	58.91 ± 27.1	55.87 ± 14.4	91.05	120.83	114.41	107.76
	p value	0.828#	0.771#	0.464+	0.168+	0.697#	0.208#	0.804#	0.173#
Place of residence	Muscat	58.41 ± 23.3	106.71	56.71 ± 29.7	52.47 ± 16.1	102.06	105.50	60.56 ± 20.4	99.51
	Ash Sharqiyah	54.00 ± 27.2	112.65	58.10 ± 23.5	54.04 ± 19.1	104.42	107.68	57.69 ± 20.0	80.76
	Dhofar	80.00	119.50	88.75 ± 15.9	67.50 ± 3.5	168.50	170.50	66.00 ± 19.8	183.00
	Ad Dhahirah	56.88 ± 29.8	109.50	42.78 ± 27.2	51.25 ± 5.8	103.39	113.61	53.50 ± 14.8	122.78
	Ad Dhakhiliyah	55.61 ± 23.0	96.72	49.29 ± 27.6	52.75 ± 18.5	99.89	108.91	63.00 ± 17.4	114.44
	Al Buraimi	35.00 ± 7.1	90.50	56.25 ± 32.6	52.50 ± 15.5	87.63	62.25	61.00 ± 17.4	98.25
	Al Batinah	62.64 ± 20.0	100.73	23.7	55.00 ± 16.8	106.27	106.97	60.34 ± 19.3	110.19
	p value	0.597+	0.927#	0.077+	0.947+	0.781#	0.546#	0.704+	0.092#

Marital status	Single	116.41	104.69	109.34	106.36	114.95	115.36	110.71	113.35
	Married	97.11	105.97	110.19	112.49	96.12	104.15	105.72	101.25
	Divorced	122.75	64.75	76.25	28.00	96.75	105.25	169.00	94.75
	Widowed	12.50	-	-	-	125.00	89.00	-	-
	p value	0.052#	0.629#	0.749#	0.142#	0.140#	0.571#	0.327#	0.336#
Education level	Elementary	45.00 ± 42.4	56.00	45.00	57.50 ± 0.0	82.25	89.00	48.00 ± 5.7	11.00
	Middle school	45.00 ± 23.8	84.60	51.43 ± 17.1	40.83 ± 39.2	114.50	102.71	57.14 ± 11.9	110.75
	High school	56.70 ± 20.8	97.67	49.91 ± 21.8	51.45 ± 29.3	97.88	93.63	57.67 ± 20.0	92.26
	Diploma	53.13 ± 20.8	108.06	59.01 ± 15.4	53.57 ± 28.5	91.54	105.75	58.93 ± 17.6	109.79
	Bachelor	60.97 ± 25.5	108.07	59.45 ± 20.6	54.47 ± 25.7	111.12	116.73	62.23 ± 20.3	115.45
	Postgraduate	62.67 ± 21.9	97.67	44.67 ± 19.7	53.75 ± 24.5	114.60	123.93	62.40 ± 12.7	89.50
	p value	0.241+	0.771#	0.167+	0.306+	0.431#	0.242#	0.329+	0.109#
Employment status	Student	118.71	102.01	124.43	116.65	126.61	121.73	63.89 ± 22.3	130.82
	Employed	95.32	105.62	108.25	108.10	97.19	111.64	59.94 ± 18.9	93.36
	Self-employed	101.25	96.22	73.56	111.67	104.06	108.56	54.67 ± 20.3	76.39
	Unemployed	92.34	91.70	90.81	95.92	90.08	87.93	57.76 ± 16.6	91.37
	Seeking a job	116.67	106.54	110.79	102.44	106.96	103.40	60.08 ± 18.4	116.25
	Retired	104.79	117.67	115.38	126.63	88.59	120.50	63.00 ± 17.8	110.75
	p value	0.222#	0.814#	0.148#	0.622#	0.092#	0.214#	0.796+	**0.009#**

SD = standard deviation.

**Assessed using the 36-Item Short-Form Health Survey. †SD values could not be calculated for variables that did not follow normal distribution. ‡Calculated using a Mann-Whitney U test for non-normally distributed variables with 2 categories. #Calculated using a Kruskal Willis test for non-normally distributed variables with ≥3 categories. +Calculated using an analysis of variance test for normally distributed variables with ≥3 categories..*

Table 5: Associations between selected clinical and social variables and health-related quality of life domain scores* among Omani adult patients with sickle cell disease (N = 235).

Variable		HRQoL domain, Mean score ± SD† or mean rank							
		Physical functioning	**Energy/fatigue**	**Pain**	**General health**	**Role limitations due to physical health**	**Role limitations due to emotional problems**	**Emotional wellbeing**	**Social functioning**
Complications (n)	1–3	102.26	50.92 ± 16.2	109.06	105.00	100.08	107.17	60.75 ± 18.3	107.18
	4–6	97.35	47.73 ± 19.8	94.16	97.57	95.72	91.47	57.82 ± 20.4	89.94
	≥7	57.53	40.94 ± 25.7	61.11	65.38	61.92	73.50	55.53 ± 15.8	66.31
	p value	**0.005#**	**0.097+**	**0.003#**	**0.026#**	**0.014#**	**0.018#**	0.417+	**0.006#**
Hx of pregnancy difficulties	Yes	48.86 ± 19.6	43.57	40.70	42.92	43.22	46.82	59.08 ± 18.9	44.40
	No	59.46 ± 27.1	46.61	63.52	59.55	55.77	52.38	59.86 ± 19.9	53.35
	p value	0.069×	0.603‡	**<0.001‡**	**0.007‡**	**0.032‡**	0.341‡	0.855×	0.138‡
Hx of exchange transfusions	Yes	95.19	95.65	93.96	99.72	94.61	100.75	106.00	97.77
	No	114.70	113.41	123.68	116.93	113.48	116.18	110.74	114.29
	p value	**0.020‡**	**0.033‡**	**<0.001‡**	0.043‡	**0.019‡**	0.056‡	0.577‡	0.046‡
Hx of blood transfusions	Yes	106.47	104.04	101.48	104.91	101.94	109.68	108.44	107.32
	No	103.33	105.61	126.58	116.66	108.71	105.83	106.99	102.76
	p value	0.728‡	0.864‡	0.007‡	0.201‡	0.438‡	0.659‡	0.875‡	0.616‡
Recent‖ ED visits/hospital admissions due to SCD	0	66.62 ± 23.5	54.41 ± 18.1	146.27	59.37 ± 16.8	117.39	118.31	67.45 ± 17.3	129.82
	1–2	56.39 ± 19.6	51.14 ± 17.6	92.65	52.81 ± 15.2	95.01	104.30	60.05 ± 19.2	93.55
	3–4	51.20 ± 22.7	42.69 ± 19.9	61.95	45.71 ± 18.1	79.70	73.96	53.57 ± 20.7	63.98
	5–6	48.33 ± 18.1	37.50 ± 17.5	61.33	36.67 ± 17.8	64.00	55.00	43.33 ± 18.8	83.00

	≥7	46.39 ± 25.9	47.37 ±17.7	48.50	44.17 ± 12.4	73.83	90.69	54.12 ± 12.2	74.55
	p value	**<0.001+**	**0.024+**	**<0.001#**	**<0.001+**	**0.001#**	**0.001#**	**<0.001+**	**<0.001#**
Compliant with medications	Yes	56.11 ± 23.0	95.82	95.82	100.37	95.82	99.12	59.45 ± 19.6	58.37
	No	59.44 ± 23.7	102.46	115.87	105.10	100.84	107.71	60.91 ± 18.6	101.71
	p value	0.337ˣ	0.438‡	**0.021‡**	0.583‡	0.542‡	0.299‡	0.609ˣ	0.692‡
Impact on academic performance	Yes	99.16	94.92	90.18	93.49	92.81	97.14	100.01	94.53
	No	111.76	117.65	139.94	132.34	119.86	124.71	117.81	122.93
	p value	0.148‡	**0.009‡**	**<0.001‡**	**<0.001‡**	**0.001‡**	**0.001‡**	**0.045‡**	**0.001‡**
Impact on social life	Yes	96.39	96.38	91.17	93.38	91.57	96.66	100.28	92.96
	No	119.81	117.34	139.41	133.14	124.57	127.59	119.37	127.92
	p value	**0.007‡**	**0.016‡**	**<0.001‡**	**<0.001‡**	**<0.001‡**	**<0.001‡**	**0.030‡**	**<0.001‡**
Hx of being bullied/belittled	Yes	100.60	88.45	92.86	88.02	86.78	87.95	85.57	90.41
	No	106.61	111.94	116.28	118.01	112.56	118.49	118.89	113.51
	p value	0.491‡	**0.006‡**	**0.008‡**	**0.001‡**	**0.002‡**	**<0.001‡**	**<0.001‡**	**0.007‡**
Family support	Yes	104.70	102.23	100.93	102.73	100.80	106.88	108.24	103.99
	No	98.66	100.94	130.44	119.17	104.23	102.45	94.52	101.48
	p value	0.560‡	0.905‡	**0.006‡**	0.117‡	0.728‡	0.657‡	0.190‡	0.807‡
Exercise frequency (no. per week)	0	95.03	99.73	102.84	51.59 ± 16.5	96.89	107.00	109.70	103.02
	1	128.40	111.15	117.42	56.72 ± 16.7	117.83	122.81	101.16	110.20
	2	112.94	103.59	108.70	55.28 ± 18.4	113.05	106.71	107.85	106.89
	≥3	114.56	122.43	130.72	54.20 ± 15.2	109.30	100.72	107.48	114.90
	p value	**0.031#**	0.360#	0.183#	0.465+	0.209#	0.501#	0.930#	0.803#

SD = standard deviation; Hx = history; ED = emergency department; SCD = sickle cell disease.

**Assessed using the 36-Item Short-Form Health Survey. †SD values could not be calculated for variables that did not follow normal distribution. ‡Calculated using a Mann-Whitney U test for non-normally distributed variables with 2 categories. #Calculated using a Kruskal Willis test for non-normally distributed variables with ≥3 categories. +Calculated using an analysis of variance test for normally distributed variables with ≥3 categories. ˣCalculated using an independent sample t-test for normally distributed variables with 2 categories. ‖Within the past 3 months.*