Research Article

Open Access Journal of
Biomedical Science
ISSN: 2690-487X

Assessment of Health-Related Quality of Life in Patients with Transfusion-Dependent Thalassemia: A Cross-Sectional Study in the United Arab Emirates

Shaikha Alshamsi[1]*, Samer Hamidi[2] and Hacer Ozgen Narci[3]

[1]*School of Health and Environmental Studies, Hamdan bin Mohammed Smart University, Dubai, UAE, ORCID: 0000-0002-1981-4544*

[2]*School of Health and Environment Studies, Hamdan Bin Mohammed Smart University, Dubai, UAE*

[3]*Department of Health Management, Istinye University, Istanbul, Turkey*

ABSTRACT

Background: Patients with transfusion-dependent thalassemia (TDT) have experienced an improvement in life expectancy due to improved therapy. Quality-of-life of these patients has become a significant component of care, and only a few studies have reported health-related quality of life (HRQoL) in adult patients with TDT compared to pediatric patients with TDT. Therefore, this study aimed to assess the HRQoL of TDT adult patients and identify the risk factors for poor HRQoL in TDT adult patients.

Methods: A descriptive cross-sectional study included 187 TDT adult patients at Dubai thalassemia center in whom HRQoL was assessed using the short-form health survey (SF-36). Descriptive analysis was done using median and interquartile range, and non-parametric tests were used for comparison of continuous variables. Regression techniques were used to study the factors affect the HRQoL among TDT adult patients. Data analysis was performed using STATA BE17.

Results: We found that patient age, employment status, and hepatic complications were negative predictors of the physical health composite (PHC) score, whereas patients' educational levels were positive predictors of the PHC score. The patient's age and employment status were negative predictors of the mental health composite (MHC) score, whereas their gender and monthly income were positive predictors of the MHC score. Moreover, we found that increasing patients' age, unemployment, treatment with combined iron-chelation therapy, and hepatic complications were associated with an increased risk factor for impaired PHC score. Being male, married, expatriate, and having a college-level education were associated with an increased risk factor for impaired MHC scores.

Conclusions: For developing multidimensional strategies for different aspects of the disease, such as training programs and psychological support, regular interviews focusing on older women and patients with lower educational levels should be considered. Furthermore, promotion of knowledge regarding the benefits of therapeutic and prophylactic processes may reduce distress and increase the quality of therapy.

ABBREVIATIONS: TDT: Transfusion-dependent thalassemia; UAE: United Arab Emirates; QoL: Quality of Life; HRQoL: Health-related quality of life; ICT: Iron Chelation Therapy; OR: Odds ratio. 36-item short form health survey (SF36).

Quick Response Code:

Address for correspondence: Shaikha Alshamsi, Hamdan bin Mohammed Smart University, UAE

Received: December 06, 2022 **Published:** January 11, 2023

How to cite this article: Shaikha A, Samer H, Hacer Ozgen N. Assessment of Health-Related Quality of Life in Patients with Transfusion-Dependent Thalassemia: A Cross-Sectional Study in the United Arab Emirates. 2022- 4(6) OAJBS.ID.000533. **DOI:** 10.38125/OAJBS.000533

INTRODUCTION

Transfusion-dependent thalassemia (TDT) is one of the most widespread inherited diseases around the world and there were approximately 399 million thalassemia carriers in 2019 [1]. Patients with TDT require lifelong, regular blood transfusions and iron chelation therapy (ICT). Lifelong transfusion and iron overload are associated with risks and complications including progressive multiorgan failure [2]. The quality-of-life (QoL) of patients with TDT is the main index of treatment effectiveness [3]; monitoring the QoL is important for detecting any impairment and initiating appropriate interventions. Hemoglobinopathies are among the most frequent monogenetic disorders in the United Arab Emirates (UAE) [4, 5]. □-thalassemia constitutes a significant health problem in the UAE because most □-thalassemia mutations are severe [6]. Currently, the number of thalassemia major patients in the UAE is decreasing, whereas the thalassemia carrier population has gradually increased [7] because of marriages between carriers [7].

Over the past three decades, regular blood transfusion and ICT have significantly enhanced the QoL of TDT patients [8]. Advanced medical management has increased patients' lifespan; however, increasing complications of the disease are expected to negatively affect patients' QoL [9]. Although patient survival is a recommended outcome, TDT patients' QoL is essential; mental and social functioning are constraints in TDT [10,11]. The QoL of TDT patients should be a significant indicator of effective therapy [12]. Many studies are conducted worldwide for investigating health-related quality of life (HRQoL) among patients with TDT to draw public attention and provide a better living environment for patients. Previous studies have shown that TDT patients have relatively worse QoL than the healthy population [13], and lower QoL scores were correlated with lower income, poor compliance with ICT, and high serum ferritin levels; presence of complications such as cardiac, hepatic, and psychiatric problems were reported as determinants of worse QoL scores; however, age, higher educational levels, lower ferritin levels, and oral ICT were associated with better QoL scores [14-19]. Well-educated respondents had lower scores during emotional and physical distress [20]. The employment status of TDT patients has an essential impact on QoL, which causes many negative health effects; the unemployment status leads to reduced QoL and worsened mental health [21]. Although QoL has become a significant component of care [22], only one study was performed in the UAE on QoL in adult patients with TDT [23]. Thus, we aimed to evaluate the HRQoL of adult patients, identify the factors affecting HRQoL, and identify the risk factors for poor HRQoL to aid decision-makers in developing clinical, counselling, and social support programs that could positively affect treatment outcomes.

MATERIALS AND METHODS

A descriptive cross-sectional study design was used to evaluate the HRQoL of adult patients with TDT, identify the factors affecting HRQoL, and identify the risk factors for poor HRQoL. The Dubai Thalassemia Center has approximately 850 patients; among these, around 450 (52.9%) are on a regular blood transfusion [24]. We included all patients aged >18 years who were diagnosed with TDT; patients with mental disorders or language difficulties were excluded. Sample size was calculated based on the following formula [25],

$$n = (P[1-P]/ (A2/Z2) + P[1-P]/N)/R \quad (1)$$

Where:

n = sample size required

N = number of people in the population (332 adults)

P = estimated variance in the population

A = Precision desired

Z = Based on confidence interval

R = Estimated response rate

According to the previous equation (1) and based on the 5% error margin with a 95% level of confidence, a minimum of 178 TDT patients were required to conduct this study. Considering that 5% participants would be excluded from the analysis because of possible non-responses, 187 of them were interviewed using a random sampling technique.

The Short Form Health Survey (SF-36) was used to conduct this study. Information regarding sociodemographic characteristics was collected by interviewing patients, whereas clinical characteristic information was extracted from their medical records after receiving ethical approvals. The reliability and validity of the SF-36 is well-documented in different languages [26-29]. Two pilot-tested versions of Arabic and English questionnaires were used in this study. Informed consent was obtained from all participants after explaining the study design and objectives. All research was performed in accordance with the ethical standards of the Research Ethics Committee at Hamdan Bin Mohammed Smart University and Dubai Scientific Research Ethics Committee, and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. All analyses were carried out in anonymous format.

Descriptive analysis was performed using median and interquartile ranges for skewed data. The Mann–Whitney U test and Kruskal–Wallis H test were used for comparison of variables. The Spearman's correlation coefficient was used to study the association between continuous variables. A linear regression model was used to identify the factors of HRQoL, and logistic regression was used to study the risk factors for low HRQoL scores. All inferential tests were two-sided; they utilized a 95% significance level. Statistical significance threshold of < 0.05 was used. Data analysis was performed using Stata/BE 17.0.

RESULTS

As indicated in Table 1, 187 TDT patients participated in this study. Their mean age was 30 years; the highest proportion of patients was non-UAE nationals (68.4 %). The primary type of thalassemia disease was β-thalassemia major (92 %) Figure 1; deferasirox was the most common ICT used (66.8%) (Table 1).

The median total score of the physical health composite (PHC) was 61.9, and the median total score of the mental health composite (MHC) was 58.8. The highest median score in the PHC was in the physical function (PF) subscale 85.0, and the lowest median score was in role limitations due to physical function (RP) 0.0. For the MHC, the highest median score was in mental health (MH) 72.0, while the lowest was in Role limitations due to mental health (RE) 33.3; (Figure 2).

The median PHC score was higher in patients with college-level education than in those with school-level and no education

(p= 0.031). Similarly, patients with the college-level education had a higher MHC median score compared to those with school-level and no education (p= 0.019). Employed patients had higher median PHC and MHC scores than unemployed patients (p= 0.027 and 0.005, respectively). Moreover, patients from the UAE had a higher median MHC score than those not from the UAE (p= 0.013). Patients with a higher number of complications (>2) had a lower median PHC score (p= 0.026) (Table 2).

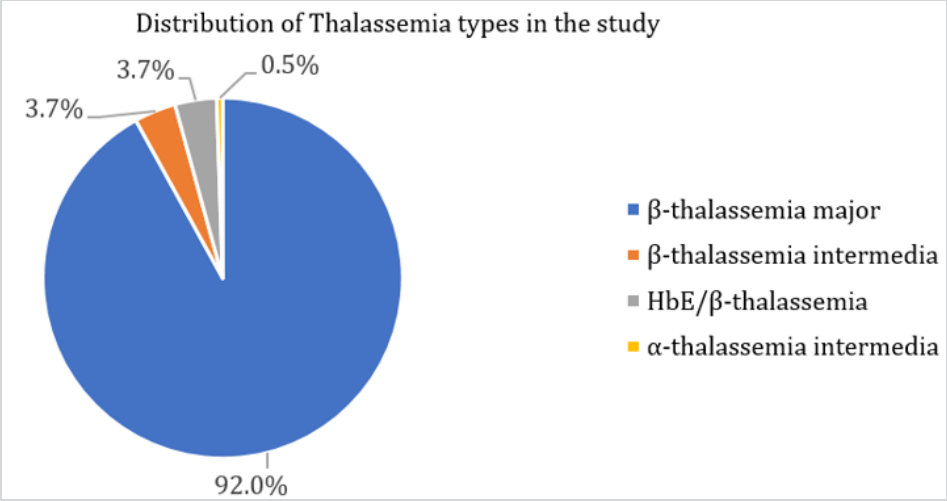

Figure 1: Distribution of transfusion-dependent thalassemia types in the study.

Table 1: Demographic and clinical characteristics of patients.

Characteristics	*N*(%)
Age in years (mean, SD) Age (*n*,%)	29.6 (6.5%)
19-35	151 (80.7%)
36-52	36 (19.3%)
Gender	
Male	97 (51.9%)
Female	90 (48.1%)
Education Level	
No formal education	7 (3.7%)
School-level	109 (58.3%)
Graduate-level	71 (38.0%)
Marital Status	
Single	126 (67.4%)
Married	59 (31.5%)
Divorced	2 (1.1%)
Employment Status	
Employed	87 (46.5%)
Unemployed	100 (53.5%)
Household Monthly Income (AED)	
< 5000	74 (39.6%)
5,000 - 9,999	34 (18.2%)
10,000 - 19,999	43 (23.0%)
20,000 - 29,999	25 (13.4%)
30,000 - 39,999	5 (2.7%)
≥ 40,000	6 (3.2%)

Nationality	
UAE	59 (31.6%)
Non-UAE	128 (6.4%)
Disease Type	
β-thalassemia major	172 (92.0%)
β-thalassemia intermedia	7 (3.7%)
HbE/β-thalassemia	7 (3.7%)
α-thalassemia intermedia	1 (0.5%)
Type of Chelation	
Deferasirox	125 (66.8%)
Deferiprone	10 (5.3%)
Deferoxamine	26 (13.9%)
Combined therapy	26 (13.9%)
Ferritin Level	
<2000 ng/ml	70 (37.4%)
≥2000 ng/ml	117 (62.6%)
History of Splenectomy	
No	162 (86.6%)
Yes	25 (13.4%)
Complication Status	
No	5 (2.7%)
Yes	182 (97.3%)
Complications no.	
≤2 complications	152 (81.3%)
>2 complications	30 (16.0%)
Cardiac Complications	
No	169 (90.4%)
Yes	18 (9.6%)
Hepatic Complications	
No	146 (78.1%)
Yes	41 (21.9%)
Spleen Complications	
No	180 (96.3%)
Yes	7 (3.7%)
Endocrine System Complications	
No	85 (45.5%)
Yes	102 (54.5%)
Bone Complications	
No	35 (18.7%)
Yes	152 (81.3%)

Table Abbreviations: AED: United Arab Emirates Currency (Dirham).

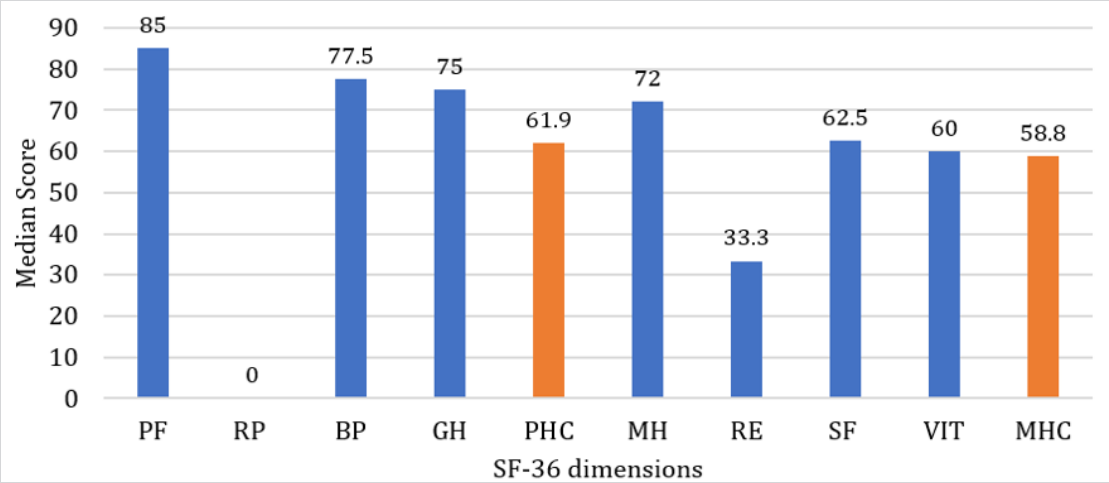

Figure 2: Median scores of the SF-36 dimensions.

Abbreviations: PF: Physical functioning; RP: role limitations due to physical problems; BP: bodily pain, GH: general health; PHC: physical health composite; MH: metal health; RE: role limitation due to emotional problems; SF: social functioning; VIT: vitality; MHC: mental health composite.

Table 2: Distribution of the physical health composite (PHC) and the mental health composite (MHC) among TDT adult patients' demographic, socioeconomic and clinical characteristics (*N*=187).

Item	PHC		MHC	
	Median(IQR)	*p*-Value	Median (IQR)	*p*-Value
Gender				
Male (*N*=97)	61.3 (20.6)	0.768	51.8 (32.0)	0.163
Female (*N*=90)	64.06 (30.3)		62.0 (32.1)	
Nationality				
UAE (*N*=59)	66.3 (30.6)	0.089	63.9 (33.0)	0.013
Non-UAE (128)	61.3 (23.3)		54.5 (35.8)	
Marital Status				
Single (*N*=126)	61.9 (23.8)	0.486	59.5 (34.0)	0.407
Married (*N*=59)	62.5 (31.3)		50.8 (28.4)	
Divorced (*N*=2)	71.25 (-)		74.3 (-)	
Educational Level				
No formal education (*N*=7)	57.5 (15.0)	0.031	44.5 (20.9)	0.019
School-level (*N*=109)	58.8 (22.5)		54.4 (30.5)	
College-level (*N*=71)	66.9 (28.1)		65.1 (33.5)	
Employment Status				
Employed (*N*=87)	63.1 (30.6)	0.027	65.1 (36.0)	0.005
Unemployed (*N*=100)	61.3 (22.3)		52.8 (30.8)	
Ferritin Level				
<2000 ng/ml (*N*=70)	62.8 (27.0)	0.429	58.8 (36.9)	0.665
≥2000 ng/ml (*N*= 117)	61.4 (26.3)		58.6 (28.5)	

Splenectomy				
No (*N*= 162)	66.3 (31.3)	0.147	54.4 (42.1)	0.477
Yes (*N*= 25)	61.6 (24.7)		59.0 (33.5)	
ICT				
Deferasirox (*N*= 125)	62.5 (26.1)	0.7	58.1 (33.8)	0.291
Deferiprone (*N*=10)	58.1 (16.7)		63.8 (41.8)	
Deferoxamine (*N*= 26)	63.1 (24.7)		66.0 (33.2)	
Combined treatment (*N*= 26)	58.1 (26.9)		55.6 (31.6)	
Number of Complications				
≤2complications (*N*-157)	62.5 (25.3)	0.026	60.3(34.3)	0.695
>2 complications (*N*=30)	57.7 (28.1)		51.7(29.0)	

Table Abbreviations: PHC: Physical health composite; MHC: Mental health composite; ICT: iron-chelation therapy; ICT: iron-chelation therapy; IQR: interquartile range.

The median PHC score showed a significantly weak negative correlation with patient age (p= 0.02). In contrast, the median MHC score showed a significantly weak positive correlation with monthly income (p= 0.001) (Table 3).

Table 3: The association between the PHC and MHC score with Patients' age and monthly income.

Item	Spearman's Coefficient (ρ)	*P*-Value
PHC (*N*=178)		
Age, Years	-0.167	0.021
MHC (*N*=178)		
Monthly Income (AED)	0.237	0.001

Table Abbreviations: PHC: Physical health composite; MHC: Mental health composite; AED: United Arab Emirates Currency (Dirham).

Male patients reported significantly higher median scores for physical functioning than female patients (p= 0.011). Patients from the UAE had significantly higher median scores in general health (p= 0.001) and physical functioning (p= 0.008) than those not from the UAE. Single patients reported a significantly higher median score for physical functioning than married or divorced patients (p= 0.049). In comparison, divorced patients reported a significantly higher median score for bodily pain than single and married patients (p= 0.020). Employed patients showed more evidence regarding role limitations due to physical problems than unemployed patients (p= 0.009). Among different iron chelators, patients treated with deferasirox showed a higher median score for body pain than those treated with other iron chelators (p=0.007). Patients with more complications showed a lower median physical functioning score (p=0.003) (Table 4).

Table 4: The distribution of the PHC subscales among TDT patients' demographic and clinical characteristics.

Item	GH		PF		RP		BP	
	Median	*P*-value*	Median	*P*-value	Median	*P*-value	Median	*P*-value
Gender								
Male (*N*=97)	60	0.546	90	0.011	0	0.181	77.5	0.068
Female (*N*=90)	65		80		25		76.25	
Nationality								
UAE (*N*=59)	70	0.001	90	0.008	0	0.745	77.5	0.734
Non-UAE (128)	60		82.5		0		77.5	

Marital Status								
Single (*N*=126)	62.5	0.751	85	0.049	0	0.47	77.5	0.02
Married (*N*=59)	65		80		25		67.5	
Divorced (*N*=2)	67.5		65		62.5		90	
Educational Level								
Illiterate (*N*=7)	55	0.78	85	0.939	0	0.005	90	0.435
School-level (*N*=109)	65		85		0		77.5	
College level (*N*=71)	65		85		50		77.5	
Employment Status								
Employed (*N*=87)	65	0.064	90	0.33	25	0.009	77.5	0.965
Unemployed (*N*=100)	60		85		0		77.5	
ICT								
Deferasirox (*N*=125)	62.5	0.636	85	0.051	0	0.572	78.8	0.006
Deferiprone (*N*=10)	57.5		72.5		37.5		62.5	
Deferoxamine (*N*=26)	65		82.5		50		67.5	
Combined treatment (*N*=26)	65		90		0		67.5	

Table Abbreviations: GH: General health; PF: Physical function; RP: Role limitations due to physical function; BP: Bodily pain; ICT: Iron-chelation therapy.

*Mann Whitney U test was used for dichotomous variables, and Kruskal Wallis was used for multinomial variables.

The median score for general health was significantly weakly negatively correlated with patient age ($p< 0.001$). The median physical functioning score was significantly moderately negatively correlated with the patient age ($p< 0.001$). The median score for bodily pain was significantly weakly negatively correlated with patient age ($p= 0.048$) (Table 5).

Table 5: The distribution between the PHC subscales and patients' age, and monthly income (*N*=187).

Item	Age		Monthly Income	
	ρ-Value*	*P*-Value	*ρ*-Value	*P*-Value
General health (GH)	-0.259	<0.001	0.17	0.116
Physical functioning (PF)	-0.363	<0.001	0.096	0.378
Bodily pain	-0.145	0.048	0.048	0.622

*ρ: rho coefficient (Spearman's rank order correlation).

Employed patients achieved higher median scores for social functioning ($p= 0.038$), role limitation due to emotional problems ($p= 0.005$), and social functioning ($p= 0.032$) than unemployed patients. Patients from the UAE had significantly higher median vitality scores ($p= 0.015$) and mental health scores ($p=0001$) than those not from the UAE. Patients with college-level education reported higher median scores for role limitation due to mental problems ($p= 0.015$) and social functioning ($p= 0.007$) compared to patients with school-level and no education (Table 6).

The median vitality score was significantly and weakly negatively correlated with monthly income ($p< 0.001$). The median mental health score was significantly, weakly, and negatively correlated with patients' monthly income ($p= 0.017$) (Table 7).

Identifying Predictors of the PHC and MHC among TDT Patients

One-year increase in patient age decreased the PHC score by 0.62. Moreover, unemployed patients were expected to have a 7.4 lower score in the PHC than employed patients. Patients with college-level education would have a greater score (15.0) than those with school-level and no education. Patients with hepatic complications were expected to have a decrease in the PHC score by 6.3 compared to those without hepatic complications, while holding all other variables in the model constant. The model indicated that

15.1 % of the MHC score variance could be predicted from the patients' employment status (p= 0.002). Unemployed patients were expected to have a 9.19 lower MHC score than employed patients. A one-year increase in patient age would yield a 0.58 lower MHC score. Female patients were expected to have a 5.66 greater MHC score than male patients. A 1% increase in the monthly income would result in a 0.03% increase in the MHC scores (Table 8).

Table 6: The distribution of the MHC subscales among TDT patients' demographic and clinical characteristics.

Item	RE		VIT		MH		SF	
	Median	*P*-Value*	Median	*P*-Value	Median	*P*-Value	Median	*P*-Value
Nationality								
UAE (*N*=59)	74	0.05	65	0.015	80	0.001	75	0.773
Non-UAE (128)	0		60		70		62.5	
Educational level								
Illiterate (*N*=7)	0	0.015	60	0.988	68	0.837	50	0.007
School-level (*N*=109)	0		60		72		62.5	
College-level (*N*=71)	66.8		60		72		75	
Employment status								
Employed (*N*=87)	66.7	0.038	65	0.072	76	0.005	75	0.032
Unemployed (*N*=100)	0		60		68		62.5	

Table Abbreviations: RE: role limitations due to emotional problems, VIT: vitality, MH: mental health, SF: social function.
*Mann Whitney U test was used for dichotomous variables, and Kruskal Wallis was used for multinomial variables.

Table 7: The correlation between the MHC subscales and TDT patients' age and monthly income.

Item	Age		Monthly Income	
	ρ-Value*	*P*-Value	*ρ*-Value	*P*-Value
Vitality (VIT)	0.087	0.424	-0.275	<0.001
Mental Health (MH)	0.183	0.091	-0.175	0.017

*ρ: rho coefficient (Spearman's rank order correlation)

Table 8: Factors associated with the PHC and MHC among patients.

Parameter	Coef.	Std. Err	P>|z|	95% CI
Physical Health Composite (PHC)				
Age	-0.6	0.2	0.003	-1.02–(-0.22)
Educational level				
School-level	8.9	7	0.203	-4.86–22.70
College-level (ref. illiterate)	15	7.2	0.038	0.83–29.1
Employment status				
Unemployed	-7.4	2.6	0.005	-12.5–(-2.2)
Hepatic complications				
Yes	-6.3	3.2	0.047	-12.5–(-0.07)
Mental Health Composite (MHC)				
Gender				
Female	5.7	2.8	0.048	0.05–11.3
Age	-0.6	0.2	0.012	-1.0–(-0.1)
Employment status				
Unemployed	-9.2	3	0.002	-15.1–3.3
Monthly income	3.2	1.5	0.035	0.2–6.3

Risk Factors for Low HRQoL in TDT Patients

Patients were stratified into two groups according to the physical and mental health composite with a cut-off of 50 points [30]; the relationship between the PHC and MHC, with potential risk factors, was explored. Patients who were unemployed (odds ratio [OR]: 2.40, *p*=0.029), were older in age (OR: 1.10, *p*=0.006), had hepatic complications (OR: 3.12, *p*= 0.009), and underwent treatment with combined therapy (OR:4.00, p= 0.008) were at a higher risk of low PHC score when all other variables in the model were constant. For MHC score, male (OR: 2.16, *p*=0.019), married (OR: 2.26, *p*=0.022), and expatriate patients (OR:2.28, *p*=0.025) had a significantly higher risk of low MHC. Patients with a college-level education had a significantly lower risk factor for low MHC (OR: 0.08, *p*=0.028) than illiterate patients when all other variables were constant (Table 9).

Table 9: Logistic regression analysis of patients with a PHC < 50 and MHC < 50.

Parameter	Odds Ratio	Std. Err	*P*>|z|	95% CI
Physical Health Composite (PHC<50)				
Age	1.1	0.03	0.003	1.03–1.17
Employment status				
Unemployed	2.4	0.96	0.029	1.10–5.26
ICT				
Deferiprone	0.26	0.3	0.243	0.03–2.47
Deferoxamine	1	0.55	0.986	0.35–2.95
Combined treatment (ref. Deferasirox)	4	2.08	0.008	1.45–11.07
Hepatic complication				
Yes	3.12	1.36	0.009	1.33–7.32
Mental Health Composite (MHC<50)				
Gender				
Male	2.16	0.71	0.019	1.13–4.10
Nationality				
Non-UAE	2.28	0.84	0.025	1.11–4.70
Marital Status				
Married (ref. single)	2.26	0.81	0.022	1.12–4.54
Educational level				
School-level	0.17	0.2	0.122	0.08–2.56
College-level (ref. illiterate)	0.08	0.09	0.028	0.01–0.75

Note: ICT: Iron Chelation Therapy.

DISCUSSION

Many studies are conducted worldwide to investigate HRQoL among TDT patients; only a few studies have reported HRQoL in adult patients compared to pediatric patients with TDT. We focused on HRQoL outcomes in adult TDT patients. Most studies reported low scores on all HRQoL subscales [13,15,31-34]. These results are consistent with our results; the median score of PHC was 61.9, and the median score of MHC was 58.8. Similarly, role limitation due to physical and emotional functions was the most affected domains in TDT patients' QoL [23]. These low scores could be due to disease complications, repeated visits to hospitals, poor management of iron-overload, shortage of psychosocial support, infertility, and knowingly short lifespan instead of all possible management measures [35].

In our study, the median physical function score was 85.0, which was the highest among the SF-36 scores. Like the study conducted by Firouz et al. the highest score was observed for physical functioning [36]. Additionally, Safizadeh et al. [17] reported that the highest score was observed in physical and social functioning [17]. The physical functioning subscale, which was better than that in other domains, was similar between our study and previous; thus, suitable treatments would decrease patients' physical problems. A negative association was found between patient age and HRQoL subscale scores. Older patients had a lower score for physical health composite; they experienced more limitations in general health and physical functioning subscales and complained more about pain. This finding is consistent with that of a previous study, which reported that older age is associated with lower HRQoL [15, 19, 31]. This could be because of the cumulative effects of complications associated with TDT that affect QoL in older patients. In contrast to our results, Haghpanah et al. [14] reported no significant relationship between patient age and QoL scores [14].

Herein, male patients had better physical function than female patients. This finding is consistent with Jameel and Im's study, which showed that females had a lower score than males [18].

Sobota et al. [19] also showed that lower QoL scores were prelavent among women [19]. Additionally, Haghpanah et al. [14] reported that women had lower scores for bodily pain and role limitation owing to emotional function [14]. These findings may be explained by sex-related differences in coping with illness; they emphasize the importance of considering patients' characteristics in managing TDT patients in general practice. However, one study found no significant differences in all HRQoL scores between men and women [32] possibly because major problems from thalassemia are common in both sexes.

Patients from the UAE had better general health, physical function, vitality scores, mental health, and mental health composite than expatriate patients. This variation in HRQoL scores could be explained by the higher socioeconomic level, social support, and free, high-quality healthcare services in all government healthcare facilities. Single patients had better physical function than others; divorced patients were less likely to complain of pain than others. In our study, most patients aged 19–35 years were single or divorced and most older patients were married. There is a relationship between patients' marital status and HRQoL, which differs by age and sex [37].

Highly educated patients experienced fewer physical and emotional limitations, better social function, and better overall physical and mental health composite compared to illiterate patients and those with school-level education. Similarly, Tajvar et al. [38] found that higher education was associated with better QoL scores in the elderly population [38]. Ansari et al. showed that higher education was associated with better QOL scores [39]. According to Lasheras et al., lower educational levels were associated with unhappiness, poor social relationships, poor self-assessed health, and sensory problems among the elderly [40]. This is because higher awareness and education levels of patients (according to their treatment regimens and follow-up visits) positively affect their QoL. Moreover, higher education, social, and income levels positively affect HRQoL [41].

Education is an important indicator that may directly or indirectly influence HRQoL through its association with higher social class and economic status [42]. In contrast to our results, Haghpanah et al. [14] showed no significant association between education levels and SF-36 scores [14].

Employed patients experienced fewer physical and emotional limitations, better mental health and social function, and better overall physical and mental health. This was supported by other studies, which found that employed patients experienced fewer physical and mental limitations, better physical functioning, mental health, social function, and better overall physical and mental health composite compared to unemployed patients [15, 31].

Patients with higher incomes had higher vitality scores, and better overall mental health. It was previously reported that socioeconomic status is associated with higher HRQoL. Haghpanah et al. [14] reported that higher income was a significant predictor of higher QoL scores [14]. This was consistent with the results of the study by Tajvar et al. [38] which showed that TDT patients could address their disease complications efficiently with better economic levels [38].

Patients treated with deferasirox were less likely to complain of pain than those treated with other ICTs. This is consistent with the study by Safizadeh et al. [17] who showed that patients receiving oral chelators had better QoL in the social functioning and mental health domains [17]. Another supporting study found that oral iron chelators could improve QoL among TDT patients [43]; this could be explained by the effectiveness of deferasirox over other ICTs. Deferasirox is an oral ICT administered once a day; it offers effective long-term ICT management of iron overload and associated TDT complications. Additionally, patient satisfaction with deferoxamine was low, mainly because of local injection-site reactions and the therapy's constraining nature. Furthermore, when presented with hypothetical oral ICT, patients unanimously preferred oral ICT to the existing treatment [44]. Other side effects experienced by patients following deferoxamine administration include neutropenia, hematological toxicity, shortness of breath, headaches, and dizziness [45].

Disease complications including cardiac, liver, spleen, endocrine, and bone diseases, present in 97.3% of our adult patients. Our results showed that more complications were associated with worse physical functioning and physical health composite scores. Similarly, Sobota et al. [19] reported lower scores among TDT patients with more complications [19]. Ansari et al. [39] reported that cardiac disease, liver disease, and psychiatric problems were associated with lower QoL scores [39]; these complications should be managed through periodic follow-ups and examinations. TDT negatively impacted patients' emotional roles, social functioning, and mental health, emphasizing the importance of psychosocial support for patients with TDT [32]. Supporting healthy emotional functioning is essential for psychological well-being and physical health because it may impact compliance with medical regimens [46].

Most studies suggest that age is a determinant factor of TDT. Older age was associated with worse HRQoL outcomes [15,31] possibly because older patients face more TDT complications and psychosocial challenges that affect different aspects of QoL. Unemployed patients had worse physical and mental HRQoL outcomes than employed patients. Many studies revealed that employment is not a source of income only; it helps in maintaining social contact, developing certain skills, and providing rights, which affect patients' QoL [47,48]. Positive QoL is associated with high self-esteem and satisfaction during spare time [48]. Patients with higher educational degrees had better physical HRQoL scores. Monthly income positively affected mental HRQoL. Similar to our study, patients with higher educational levels had better physical functioning and mental health [20,39,49]. This could be because TDT patients with higher education get better jobs and incomes; there is overwhelming evidence that higher monthly income leads to better QoL. Hepatic complications negatively affected physical HRQoL outcomes. TDT complications such as hepatitis and cardiac disease are associated with deteriorated HRQoL [15,39]; hepatic complications lead to liver dysfunction, several endocrine problems, and recurring anemia, negatively affecting patients' QoL through reduced physical function and vitality, deteriorated general health, and increased bodily pain. Interestingly, women had better mental HRQoL outcomes than men. Surprisingly, the opposite finding was noted by Jameel and Im, where female patients showed lower scores than male patients [18]. Many studies failed to report a significant difference in HRQoL scores between males and females, indicating that disease problems are common in both sexes [31,32]. The explanation for our finding could be that female patients have higher education levels and incomes compared to male patients. Consequently, women have a better mental health status than men.

Several studies on HRQoL reported low physical and mental health scores for TDT and identified many forms of physical

health disorders related to severe anemia and iron overload. Moreover, mental health disorders, such as depression and stress, have also been reported. In this study, we determined the factors associated with decreased PHC and MHC scores. As mentioned earlier, increasing age was associated with more complications that severely impacted the PHC score. Like previous studies [47,50] we found that unemployment creates physical and mental health deterioration because financial problems, stress, and loss of social contact could lead to negative health and social consequences. Many studies revealed that employed people have better QoL than unemployed people [21,51]. Hepatic complications are associated with a high risk of poor PHC. Hepatic diseases remain one of the most serious problems among TDT [52], and chronic iron overload and viral infection may cause liver cirrhosis. Consequently, patients with liver complications experienced more physical limitations, complained of increased bodily pain, and had poor general health; therefore, they had worse PHC scores. Combined therapy was associated with a higher risk of low PHC score. In our study, more than 92% of combined therapy-treated patients had severely high ferritin levels (≥ 2000 mg/dL), and more than half of the adult patients with combined therapy were administered injectables with oral therapy. Overall, poor adherence to ICT is associated with injectable ICT [14,43] and is explained by severely high serum ferritin levels, which are associated with higher morbidities in patients with TDT. Therefore, lower ferritin levels may improve the overall QoL of patients with TDT. Decreasing disease complications should be monitored by improving therapeutic strategies to lower ferritin levels in patients with TDT [53].

In our study, men were at a higher risk of poor MHC. In contrast, Dhirar et al. [54] reported that men had a better score in the emotional domain than women [54] and Srivastava reported that women are twice as likely as men to experience mental health disorders [55]. Our result could be explained by the fact that the proportion of female patients with higher monthly income (AED >10,00) was higher than male patients, and about 47% of male patients were unemployed. Moreover, the number of male patients with cardiac, hepatic, and bone complications was higher than that of female patients, which negatively affected mental QoL. Expatriate patients are associated with a higher risk of poor MHC. As mentioned earlier, patients from the UAE have higher socioeconomic levels, family support, and free high-quality healthcare services in all government healthcare facilities. While some expatriate patients lived without their families, were unemployed, and had lower education and socioeconomic levels, others lacked resources including transportation to the Dubai Thalassemia Center. These factors can negatively affect mental health of the patient. Married patients have a higher risk of poor MHC. Likewise, another study reported that married patients experienced depression and lower anxiety than single patients [56]. In contrast, two studies revealed that married participants reported higher HRQoL than single [57] or divorced [58]. This could be because TDT patients suffer from a wide range of physical and mental problems, such as disease complications, infertility, and long-term dependence on blood transfusions, which may negatively affect their marital relationship. Higher educational levels were associated with a lower risk of poor MHC. As mentioned earlier, this could be explained by the higher socioeconomic level of patients with higher education, which positively affects the mental QoL.

This study had some limitations. First, the unavailability of some variables that could be potential predictors for the HRQoL of TDT patients, such as the age at anaemia onset and the pre-transfusion Hb level, were not extracted from patients' medical records. Second, the HRQoL scores of healthy people in the UAE were not available. Therefore, it is difficult to estimate the true magnitude of the impact of thalassemia on HRQoL.

CONCLUSION

Patients suffer from clinical and psychological disorders. Therefore, they require long-term support to prevent mental health problems. Regular psychological support should be a part of the clinical treatment plan, which can be accomplished using a multidisciplinary team-based approach. Furthermore, HRQoL could be used as evidence to introduce any additional services related to treatment adherence and psychosocial support to improve the HRQoL of TDT patients, allowing the evaluation of clinical performance and identifying any weaknesses that need to be addressed.

DECLARATIONS

Ethical Approval and Consent to Participate

The study was approved by the Research Ethics Committee at Hamdan Bin Mohammed Smart University and Dubai Scientific Research Ethics Committee – Dubai Health Authority. Informed written consent was obtained from the participants and ensuring the right to withdraw at any time during the study.

AVAILABILITY OF DATA AND MATERIAL

The data that support the findings are available from the corresponding author with permission from the Dubai Health Authority.

AUTHORS' CONTRIBUTIONS

SA, SH, and HN designed the study. Data collection, analysis, and interpretation were conducted by SA. The manuscript was drafted by SA. All authors have reviewed and approved the final manuscript.

ACKNOWLEDGMENT

We are grateful to the Dubai Thalassemia Center for their assistance during patients interviewing.

AUTHORS' INFORMATION

One Shaikha Alshamsi: School of Health and Environment Studies, Hamdan bin Mohammed Smart University, Dubai, United Arab Emirates. 2Samir Hamidi: School of Health and Environment Studies, Hamdan Bin Mohammed Smart University, Dubai, UAE. 3Hacer Ozgen Narci: Department of Health Management, Istinye University, Istanbul, Turkey.

REFERENCES

1. IMHE (2020) GBD Compare date visualization University of Washington: Institute of health metrics and evaluation.
2. Vichinsky E, Neumayr L, Trimble S, Giardina PJ, Cohen AR, et al. (2014) Transfusion complications in thalassemia patients: a report from the centers for Disease Control and Prevention (CME). Transfusion 54(4): 972-981.
3. Goli M, Salarvand S, Dehvan F, Ghafouri H, Dalvand S, et al. (2018) Health-related quality of life in Iranian patients with Thalassemia major: a systematic review and meta-analysis. International Journal of Pediatrics 6(11): 8483-8494.
4. Baysal E (2021) Hemoglobinopathies in the United Arab Emirates. Hemoglobin 25(2): 247-253.

5. Al-Gazali L, Ali BR (2010) Mutations of a country: A mutation review of single gene disorders in the United Arab Emirates (UAE). Human mutation 31(5): 505-520.

6. Baysal E (2011) Molecular basis of β-thalassemia in the United Arab Emirates. Hemoglobin. 35(5-6): 581-588.

7. Kim S, Tridane A (2017) Thalassemia in the United Arab Emirates: why it can be prevented but not eradicated. PLoS One 12(1): e0170485.

8. Telfer P, Constantinidou G, Andreou P, Christou S, Modell B (2005) Quality of life in thalassemia. Annals of the New York academy of sciences 1054(1): 273-282.

9. Bayanzay K, Alzoebie L (2016) Reducing the iron burden and improving survival in transfusion-dependent thalassemia patients: current perspectives. J Blood Med 7: 159-169.

10. Mazzone L, Battaglia L, Andreozzi F, Romeo MA, Mazzone D (2009) Emotional impact in β-thalassaemia major children following cognitive-behavioural family therapy and quality of life of caregiving mothers. Clin Pract Epidemiol Ment Health 5(1): 5.

11. Borgna-Pignatti C (2010) The life of patients with thalassemia major. Haematologica 95(3): 345-348.

12. Dahlui M, Hishamshah MI, Rahman A, Aljunid SM (2009) Quality of life in transfusion-dependent thalassaemia patients on desferrioxamine treatment. Singapore Med J 50(8): 794.

13. Töret E, Karadaş NÖ, Gökçe NÖ, Kaygusuz A, Karapınar TH, et al. (2018) Quality of life and depression in turkish patients with β-thalassemia major: A cross-sectional study. Hemoglobin 42(5-6): 326-329.

14. Haghpanah S, Nasirabadi S, Ghaffarpasand F, Karami R, Mahmoodi M, et al. (2013) Quality of life among Iranian patients with beta-thalassemia major using the SF-36 questionnaire. Sao Paulo Med J 131(3): 166-172.

15. Haghpanah S, Vahdati S, Karimi M (2017) Comparison of quality of life in patients with β-Thalassemia intermedia and β-Thalassemia major in Southern Iran. Hemoglobin 41(3): 169-174.

16. Adib-Hajbaghery M, Ahmadi M (2015) Health related quality of life, depression, anxiety and stress in patients with beta-thalassemia major. Iran J Ped Hematol Oncol 5(4): 193-205.

17. Safizadeh H, Farahmandinia Z, Nejad SS, Pourdamghan N, Araste M (2012) Quality of life in patients with thalassemia major and intermedia in Kerman Iran (I.R.). Mediterr J Hematol Infect Dis 4(1): e2012058.

18. Jameel T, Imran S (2015) The compromised quality of life in β-thalassemia major children in non- urban setup in a developing country. J Hematol Thrombo Dis 4(3): 1000245.

19. Sobota A, Yamashita R, Xu Y, Trachtenberg F, Kohlbry P, et al. (2011) Quality of life in thalassemia: a comparison of SF-36 results from the thalassemia longitudinal cohort to reported literature and the US norms. Am J Hematol 86(1): 92.

20. Ross CE, Van Willigen M (1997) Education and the subjective quality of life. J Health Soc Behav 38(3): 275-297.

21. Martella D, Maass A (2000) Unemployment and life satisfaction: The moderating role of time structure and collectivism 1. Journal of Applied Social Psychology 30(5): 1095-1108.

22. Gollo G, Savioli G, Balocco M, Venturino C, Boeri E, et al. (2013) Changes in the quality of life of people with thalassemia major between 2001 and 2009. Patient Prefer Adherence. 7: 231-236.

23. Hachim I, Al Nuaimi Y, El Baky A, Al Hashmi F, Rahman A, et al. (2014) Quality of life in patients suffering from thalassaemia in Ras Al Khaimah, United Arab Emirates. Hamdan Med J 7(1): 102-108.

24. Nuwais SA (2019) UAE records new blood disorder cases after hopes of eradicating condition. The National. Sect Health.

25. Watson J (2001) How to determine a sample size: Tipsheet# 60. University Park, PA: Penn State Cooperative Extension.

26. Brazier JE, Harper R, Jones N, O'cathain A, Thomas K, et al. (1992) Validating the SF-36 health survey questionnaire: new outcome measure for primary care. BMJ 305(6846): 160-164.

27. Sabbah I, Drouby N, Sabbah S, Retel-Rude N, Mercier M (2003) Quality of life in rural and urban populations in Lebanon using SF-36 health survey. Health Qual Life Outcomes 1(1): 1.

28. Bousquet J, Knani J, Dhivert H, Richard A, Chicoye A, et al. (1994) Quality of life in asthma. I. Internal consistency and validity of the SF-36 questionnaire. Am J Res Crit Care Med 149(2): 371-375.

29. Jenkinson C, Wright L, Coulter A (1994) Criterion validity and reliability of the SF-36 in a population sample. Qual Life Res 3(1): 7-12.

30. Kroenke CH, Kubzansky LD, Adler N, Kawachi I (2008) Prospective change in health-related quality of life and subsequent mortality among middle-aged and older women. Am J Pub Health 98(11): 2085-2091.

31. Adam S (2019) Quality of life outcomes in thalassaemia patients in Saudi Arabia: a cross-sectional study. East Mediterr Health J 25(12): 887-895.

32. Emadi Dehaghi B, Rasooli L, Mohammadi Farsani S (2016) Quality of life among patients with beta-thalassemia major in Shahrekord city, Iran. International Journal of Epidemiologic Research 3(4): 324-328.

33. Arian M, Mirmohammadkhani M, Ghorbani R, Soleimani M (2019) Health-related quality of life (HRQoL) in beta-thalassemia major (β-TM) patients assessed by 36-item short form health survey (SF-36): a meta-analysis. Qual Life Res 28(2): 321-334.

34. Porter J, Bowden DK, Economou M, Troncy J, Ganser A, et al. (2012) Health-related quality of life, treatment satisfaction, adherence and persistence in β thalassemia and myelodysplastic syndrome patients with iron overload receiving deferasirox: Results from the EPIC clinical trial. Anemia 2012: 297641.

35. Tahir H, Shahid SA, Mahmood KT (2011) Complications in thalassaemia patients receiving blood transfusion. J Biomed Sci and Res 3(1): 339-346.

36. Amani F, Fathi A, Valizadeh M, Farzaneh E, Fattahzadeh-Ardalani G (2015) Quality of life among Ardabil patients with beta-thalassemia major. Int J Res Med Sci 3(11): 3308-3312.

37. Han KT, Park EC, Kim JH, Kim SJ, Park S (2014) Is marital status associated with quality of life? Health Qual Life Outcomes 12(1): 109.

38. Tajvar M, Arab M, Montazeri A (2008) Determinants of health-related quality of life in elderly in Tehran, Iran. BMC Public Health 8: 323.

39. Ansari S, Baghersalimi A, Azarkeivan A, Nojomi M, Hassanzadeh Rad A (2014) Quality of life in patients with thalassemia major. Iran J Ped Hematol Oncol 4:57-63.

40. Lasheras C, Patterson AM, Casado C, Fernandez S (2001) Effects of education on the quality of life, diet, and cardiovascular risk factors in an elderly Spanish community population. Exp Aging Res 27(3): 257-270.

41. GuimarãesSá AM, Ferreira PAM, Souza MT, Nascimento GC, da Silva Pereira Damianse S, et al. (2018) Higher income and integration into the workforce are the main factors associated with quality of life in acromegalic patients in Northeastern Brazil. Int J Endocrinol 2018: 6135080.

42. United Nations NY (2002) NY. Department of economic, affairs S. World population ageing, United Nations Publications, pp.1950-2050.

43. Osborne RH, De Abreu Lourenço R, Dalton A, Houltram J, Dowton D, et al. (2007) Quality of life related to oral versus subcutaneous iron chelation: A time trade-off study. Value Health10(6): 451-456.

44. Abetz L, Baladi JF, Jones P, Rofail D (2006) The impact of iron overload and its treatment on quality of life: results from a literature review. Health Qual Life Outcomes 4: 73.

45. Alymara V, Bourantas D, Chaidos A, Bouranta P, Gouva M, et al. (2004) Effectiveness and safety of combined iron-chelation therapy with deferoxamine and deferiprone. Hematol J 5(6): 475-479.

46. Senol SP, Tiftik EN, Unal S, Akdeniz A, Tasdelen B (2016) Quality of life, clinical effectiveness, and satisfaction in patients with beta thalassemia major and sickle cell anaemia receiving deferasirox chelation therapy. J Basic Clin Pharm 7(2): 49.

47. Worach-Kardas H, Kostrzewski S (2014) Quality of life and health state

of long - term unemployed in older production age. Appl Res Qual Life 9(2): 335-353.

48. Axelsson L, Andersson I, Leden I, Ejlertsson G (2007) Inequalities of quality of life in unemployed young adults: A population-based questionnaire study. Int J Equity Health 6: 1.

49. Gan G, Hue Y, Sathar J (2016) Factors affecting quality of life in adult patients with thalassaemia major and intermedia. Ann Acad Med Singap 45: 520-523.

50. Jiang Y, Hesser JE (2006) Associations between health-related quality of life and demographics and health risks. Results from Rhode Island's 2002 behavioral risk factor survey. Health Qual Life Outcomes 4: 14.

51. Timperi AW, Ergas IJ, Rehkopf DH, Roh JM, Kwan ML (2013) Employment status and quality of life in recently diagnosed breast cancer survivors. Psychooncology 22(6): 1411-1420.

52. Hassan T, Zakaria M, Fathy M, Arafa M, El Gebaly S, et al. (2018) Association between genotype and disease complications in Egyptian patients with beta thalassemia: A Cross-sectional study. Scientific Reports 8(1): 17730.

53. Boonchooduang N, Louthrenoo O, Choeyprasert W, Charoenkwan P (2015) Health-related quality of life in adolescents with thalassemia. Pediatr Hematol Oncol 32(5): 341-348.

54. Dhirar N, Khandekar J, Bachani D, Mahto D (2016) Thalassemia Major: how do we improve quality of life? SpringerPlus 5(1): 1895.

55. Srivastava K (2012) Women and mental health: Psychosocial perspective. Ind Psychiatry J 21(1): 1-3.

56. Kaplan CP, Miner ME (2000) Relationships: importance for patients with cerebral tumours. Brain Inj 14(3): 251-259.

57. Porter KR, Menon U, Vick NA, Villano JL, Berbaum ML (2014) Assessment of clinical and nonclinical characteristics associated with health-related quality of life in patients with high-grade gliomas: a feasibility study. Support Care Can 22(5): 1349-1362.

58. Weitzner MA, Meyers CA, Byrne K (1996) Psychosocial functioning and quality of life in patients with primary brain tumors. J Neurosurg 84(1): 29-34.