

# Evaluation of the relationship between sociodemographic characteristics, quality of life, depression, drug compliance and biochemical parameters in patients with thalassemia major in Isparta

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## Abstract

**Aim:** We aimed to evaluate the quality of life and depression levels of BTM patients in our unit, to determine the relationship between each parameter and evaluate the demographic characteristics, laboratory data, and treatment compliance.

**Materials and Methods:** Twenty-seven BTM patients aged 18 years and over were included in the study. Sociodemographic data were collected from the patients with a data form. Laboratory data were obtained from hospital records retrospectively. The patients were asked to respond to the BDI and SF-36 content in which depression was questioned and quality of life was evaluated respectively. The relationship between each parameter was examined.

**Results:** 70,3 % of the patients had minimal-mild and 11,1 % of the patients had severe depression. Depression levels were significantly higher in patients who had a regular job and who had poor compliance to chelation therapy. Physical function and physical role subscales of single patients were significantly higher than married patients. Patients with monthly income had higher quality of life in terms of physical role, mental health, social functioning and pain subscales. The patients without co-morbidity had higher quality of life in terms of physical role, pain and general health subscales. Patients with severe depression had the worst quality of life. The general health perception of patients with minimal depression was higher than those with moderate depression and severe depression.

**Conclusions:** Psychosocial support of patients is important and, therapy should be aimed not only to ensure long life without complications but also improve the quality of life of the patients.

**Keywords:** Beta thalassemia major; depression; quality of life; treatment compliance

## INTRODUCTION

Thalassaemic syndromes are hemoglobinopathies that are characterized by deficient or absent synthesis of globin chains in haemoglobin structure and they are inherited as autosomal recessive (1). Hereditary haemoglobin disorders are the most common monogenic diseases; approximately 5.2 % of the world population carries an abnormal haemoglobin gene and it is estimated that approximately 300,000-400,000 babies are born with a severe haemoglobin disorder per year. About 17 % of these are thalassemia diseases and 83 % are sickle cell diseases (2,3). In beta thalassemia major (BTM); there is no production of  $\beta$ -globin chain as a result of homozygous or double heterozygous mutation (4).

It has the highest prevalence especially in endemic regions of malaria, including sub-Saharan Africa, the Mediterranean, Southeast Asia and the subcontinent of India (5). Approximately there are 1,300,000 thalassemia carriers and 4,500 thalassemia patients in Turkey; the prevalence of  $\beta$ -thalassemia trait is 2.1 % (6). The incidence of beta thalassemia minor was found to be 25 % among 6,054 participants in Isparta at 2002 and, the frequency of beta thalassemia was lower in the city centre than in neighbouring towns (7).

The clinics of BTM usually appear between the ages of 6 months and 2 years, and, these findings include; growth retardation, pallor, jaundice, discoloration of the skin, muscular system weakness, hepato-splenomegaly, leg

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ulcer, bone marrow enlargement and skeletal changes due to extramedullary haematopoiesis to compensate anaemia (8).

Current treatment modalities include regular blood transfusions, iron chelating therapy, splenectomy and monitoring and treatment of complications. Regular transfusion should be aimed at preventing tissue hypoxia and providing target haemoglobin values for the continuation of optimal growth (9,10). Stem cell transplantation is the only curative treatment alternative for patients with HLA compatible donors (8,11) but recently, gene therapy seems to become a viable option to patients who do not have a matched donor and do not have  $\beta^0/\beta^0$  genotype; phase 3 trial of the modality is currently ongoing (12). Uncontrolled iron overload by chronic transfusions is an important cause of mortality and morbidity by accumulating in tissues and organs and causing multiple organ damage. The most common clinical events are liver damage, heart disease and dysfunction of the endocrine glands (13,14).

Depression is an important global health problem both personally and socially, which is characterized by sadness, loss of interest and desire, not enjoying life, feeling guilty, insecurity, anorexia, sleep disorders, fatigue, and concentration disorder. According to World Health Organization (WHO) data, there are approximately 300 million patients suffering depression (15). Beck Depression Inventory (BDI) is a form created by Beck et al. in 1961 to quantitatively determine the degree of depression. While the first form needs cooperation with patient, a second form was developed in 1978 to allow individual measurement by the physician (16). In our country, validity, and reliability studies of first form were carried in 1980 and the second form in 1988 and all forms were translated to Turkish (17,18).

Quality of life is defined by WHO as perceptions of individuals' goals, expectations, standards and concerns within their culture and value systems (19). In addition to aiming the physical, mental, and social well-being of patients, reaching the patients' feelings about the disease, understanding how their illness affect their daily life and activity, becomes important in terms of general health perception. Short Form-36 (SF-36) is one of the scales designed for determining the quality of life of individuals. (20,21). It was adapted to Turkish in 1999 by Kocyigit and colleagues (22). Considering the demographic characteristics, socioeconomic and cultural variables of the Turkish society, the norm values of the Turkish society were calculated with a comprehensive study (23).

## MATERIALS and METHODS

This prospective study was approved by Ethics Committee of Suleyman Demirel University School of Medicine (05.04.2019-126). 33 patients aged 18 and over who were diagnosed and followed with BTM at Suleyman Demirel

University Department of Haematology were included in the study. After exclusion of 6 patients for insufficient data, 27 patients who received regular transfusion therapy and agreed to participate were evaluated.

Sociodemographic characteristics, medical and laboratory data of all participants were obtained from patients and medical files. Patients' age, gender, marital status, education and employment status, monthly income, comorbidity, and compliance with chelation therapy were collected. The poverty line limit has been established according to the latest data from the Statistics Agency of Turkey. Regardless of the chelator used by patients, only compliance was assessed.

The average of five pretransfusion hemogram and three ferritin parameters of the patients were calculated and used for statistics.

BDI form consists of 21 questions and each question evaluates a unique depressive symptom (24). Depression levels of patients are categorized according to total scores; minimal depression (0-9 points), mild depression (10-16 points), moderate depression (17-29 points) and severe depression (30-63 points) respectively.

The SF-36 questionnaire consists of eight subscales that evaluate the QoL (physical functioning, physical role, pain, general health, vitality, social functioning, emotional role, mental health). For each scale, the scores are transformed to 0-100 scale, with higher scores indicating better QoL. The norm values of SF-36 scale for Turkish society were determined with a study by Demiral and colleagues (23).

## Statistical Analysis

SPSS for Windows version 22.0 package program was used for statistical analysis and  $p < 0.05$  was considered statistically significant. In the case of two non-parametric groups, the Mann Whitney U test was used and more than two non-parametric groups, the Kruskal Wallis test were used. Chi square (Chi-square) tests were used to determine whether there was a significant difference in the distribution of categorical variables and between groups. Spearman Correlation analysis was used to compare SF-36 subscales and numerical data.

## RESULTS

27 patients aged 18-71 (mean  $28.2 \pm 10$ ) were included in the study. Their demographic characteristics were presented in Table 1. None of the patients had a monthly income above the poverty line. Six patients had comorbidities and, three patients did not receive chelation therapy at the time of the evaluation.

The mean haemoglobin level of the patients was 8.99 gr / dl ( $\pm 0.98$ ) and the mean ferritin level was 1849.9 ng / ml ( $\pm 726.5$ ) (Table 2).

The mean Beck depression scores of the patients were  $14.40 \pm 11.29$ . Depression scores of patients based on demographic variables were shown in Table 3.

**Table 1. The demographic characteristic of patients**

	n (%)
<b>Age</b>	
≤ 28 year	18 (% 66.7)
>28 year	9 (% 33.3)
<b>Sex</b>	
Female	11 (% 40.7)
Male	16 (% 59.3)
<b>Marital status</b>	
Married	6 (% 22.2)
Single	21 (% 77.8)
<b>Education</b>	
Primary school	9 (% 33.3)
High school	9 (% 33.3)
University	9 (% 33.3)
<b>Working status</b>	
Employed	13 (% 48.1)
Unemployed or student	14 (% 51.9)
<b>Monthly income</b>	
Unemployed	14 (% 51.9)
< poverty line	13 (% 48.1)
> poverty line	-
<b>Co-morbidity</b>	
Yes	6 (% 22.2)
No	21 (% 77.8)
<b>Compliance with chelator therapy</b>	
Never	3 (% 11.1)
Always	15 (% 55.6)
Sometimes	9 (% 33.3)

**Table 2. The mean hemoglobin and ferritin levels of patients**

	n (%)
<b>Ferritin level</b>	
<1000 ng/ml	2 (7.4)
1000-2000 ng/ml	19 (70.4)
>2000 ng/ml	6 (22.2)
<b>Hgb level</b>	
< 9 gr/dl	14 (53.9)
≥ 9 gr/dl	12 (46.1)

Beck depression score of the employed group ( $16.7 \pm 8.8$ ) was significantly higher than unemployed or student group ( $12.2 \pm 13.1$ ) ( $p=0.044$ ). When treatment compatibility was examined, a statistically significant difference was found between those who did not use chelators and those who used it irregularly ( $p=0.013$ ). No statistically significant difference was found between the ferritin and haemoglobin levels and depression levels of the patients ( $p: 0.471$  and  $p: 0.455$ , respectively). The QoL scores of the patients are shown in Table 4. The female participants had a higher score in emotional role, vitality, mental health subscales but a statistical significance were found only in the vitality subscale ( $p=0.045$ ).

**Table 3. Depression scores based on demographic variables**

	mean $\pm$ SD	P value
<b>Age</b>		
≤ 28 year	16.27 $\pm$ 12.04	0.164
>28 year	10.66 $\pm$ 9.11	
<b>Sex</b>		
Female	10.09 $\pm$ 5.20	0.126
Male	17.37 $\pm$ 13.42	
<b>Marital status</b>		
Married	8.33 $\pm$ 4.84	0.136
Single	16.14 $\pm$ 12.07	
<b>Education</b>		
Primary school	13.3 $\pm$ 16.5	0.310
High school	13.4 $\pm$ 7.1	
University	16.4 $\pm$ 9.01	
<b>Working status</b>		
Employed	16.7 $\pm$ 8.8	<b>0.044</b>
Unemployed or student	12.2 $\pm$ 13.1	
<b>Monthly income</b>		
Unemployed	12.2 $\pm$ 13.1	<b>0.044</b>
< poverty line	16.7 $\pm$ 8.8	
> poverty line		
<b>Co-morbidity</b>		
Yes	17.3 $\pm$ 6.6	0.110
No	13.5 $\pm$ 12.3	
<b>Compliance with chelator therapy</b>		
Never	4 $\pm$ 3	<b>0.013</b>
Always	12.2 $\pm$ 7.1	
Sometimes	21.4 $\pm$ 14.7	

**Table 4. The QoL scores of patients**

	Total patients mean $\pm$ SD	Female (n: 11) mean $\pm$ SD	Male (n:16) mean $\pm$ SD	P value
<b>Physical functioning</b>	82.7 $\pm$ 18.3	80.0 $\pm$ 15.0	84.6 $\pm$ 20.6	0.254
<b>Physical role</b>	60.1 $\pm$ 41.1	59.0 $\pm$ 43.6	60.9 $\pm$ 40.7	0.837
<b>Emotional role</b>	57.3 $\pm$ 43.4	57.5 $\pm$ 44.9	57.2 $\pm$ 43.8	0.876
<b>Vitality</b>	50.3 $\pm$ 19.3	60.0 $\pm$ 14.4	43.7 $\pm$ 19.7	<b>0.045</b>
<b>Mental health</b>	59.8 $\pm$ 17.8	66.9 $\pm$ 14.5	55.0 $\pm$ 18.6	0.101
<b>Social functioning</b>	67.1 $\pm$ 27.3	65.9 $\pm$ 22.4	67.9 $\pm$ 30.9	0.617
<b>Body pain</b>	66.3 $\pm$ 25.6	62.7 $\pm$ 21.3	68.9 $\pm$ 28.6	0.498
<b>General health</b>	53.8 $\pm$ 23.9	53.6 $\pm$ 22.2	54.0 $\pm$ 25.7	0.980

No significant difference was found in the overall QoL mean score of the patients with different ages and education ( $p>0.05$ ). A positive correlation was found between education levels and QoL. Table 5 also shows that single patients have higher mean scores than married patients in all subgroups of QoL. However, the differences were not significant except for physical functioning ( $p=0.012$ ) and physical role ( $p=0.048$ ). Patients who had a regular monthly income had higher scores in all QoL subscales than unemployed patients.

There was a significant difference in physical role ( $p=0.011$ ), mental health ( $p=0.016$ ), social functioning ( $p=0.037$ ) and body pain ( $p=0.046$ ) scales.

Patients with co-morbidities had lower scores in all quality of life subscales than patients without co-morbidity;

significant difference were seen in physical role ( $p=0.041$ ), general health ( $p=0.030$ ) and body pain ( $p=0.012$ ) scales. There was no significant difference between compliance with chelation therapy, haemoglobin, and ferritin levels and QoL subscales ( $p>0.05$ ) (Table 6).

Table 5. Demographic variables and QoL scales

	SF-36 scales							
	PF	PR	ER	VT	MH	SF	BP	GH
<b>Age</b>								
≤ 28 year	84.4±12.8	68.0±40.0	65.7±38.9	54.4±21.1	63.3±15.3	70.8±24.2	70.0±23.9	54.7±23.9
>28 year	79.4±26.9	44.4±41.0	40.7±49.3	42.2±12.2	52.8±21.1	59.7±32.9	59.1±28.9	52.2±25.3
<b>P value</b>	0.833	0.147	0.194	0.088	0.224	0.435	0.448	0.857
<b>Marital status</b>								
Married	65.0±30.3	29.1±40.0	55.5±50.1	41.6±14.0	56.6±24.9	54.1±30.2	49.5±26.3	42.5±18.9
Single	87.8±9.2	69.0±37.8	57.9±42.6	52.8±20.1	60.7±15.8	70.8±26.0	71.1±24.0	57.1±26.4
<b>P value</b>	<b>0.012</b>	<b>0.048</b>	0.951	0.151	0.814	0.238	0.103	0.241
<b>Education</b>								
Primary school	77.7±27.3	52.7±42.2	46.2±48.4	46.1±15.9	53.3±20.0	59.7±32.9	63.0±31.5	50.5±25.7
High school	85.0±10.0	55.5±44.6	51.8±44.4	51.1±16.5	59.1±17.2	69.4±29.3	59.4±23.5	50.0±26.3
University	85.5±14.2	72.2±38.4	74.0±36.4	53.8±25.4	67.1±14.8	72.2±19.5	76.6±20.2	61.1±20.4
<b>P value</b>	0.452	0.566	0.365	0.824	0.262	0.768	0.492	0.802
<b>Working status</b>								
Employed	86.5±12.3	80.7±29.1	71.7±38.1	54.6±19.9	67.3±13.4	78.8±18.6	76.9±21.4	61.1±19.9
Unemployed/student	79.2±22.5	41.0±42.2	44.0±45.1	46.4±18.5	52.8±18.9	56.2±30.1	56.6±26.1	47.1±26.0
<b>P value</b>	0.284	0.011	0.108	0.213	<b>0.016</b>	<b>0.037</b>	<b>0.046</b>	0.188
<b>Monthly income</b>								
Unemployed	79.2±22.5	41.0±42.2	44.0±45.1	46.4±18.5	52.8±18.9	56.2±30.1	56.6±26.1	47.1±26.0
< poverty line	86.5±12.3	80.7±29.1	71.7±38.1	54.6±19.9	67.3±13.4	78.8±18.6	76.9±21.4	61.1±19.9
<b>P value</b>	0.284	<b>0.011</b>	0.108	0.213	<b>0.016</b>	<b>0.037</b>	<b>0.046</b>	0.188

PF: physical functioning, PR: physical role, ER: emotional role, VT: Vitality, MH: Mental health, SF: Social functioning, BP: Body Pain, GH: General health

Table 6. Different variables and QoL scales

	SF-36 scales							
	PF	PR	ER	VT	MH	SF	BP	GH
<b>Co-morbidity</b>								
Yes	85.2±12.3	67.8±39.6	62.6±42.7	52.1±19.7	61.7±15.6	71.4±25.3	73.2±22.7	58.5±24.2
No	74.1±32.0	33.3±37.6	38.8±44.2	44.1±17.7	53.3±24.4	52.0±31.0	42.5±21.9	37.5±14.4
<b>P value</b>	0.402	<b>0.041</b>	0.187	0.464	0.519	0.175	<b>0.012</b>	<b>0.030</b>
<b>Compliance with chelator therapy</b>								
Never	75.0±22.9	66.6±57.7	66.6±57.7	51.6±7.6	73.3±12.2	70.8±31.4	77.5±22.5	61.6±12.5
Always	80.6±22.0	60.0±35.1	51.0±43.3	47.3±18.9	53.8±17.6	60.0±25.5	61.0±24.9	51.0±23.9
Sometimes	88.8±6.5	58.3±50.0	64.8±42.8	55.0±22.9	65.3±16.6	77.7±28.4	71.6±28.3	56.1±27.8
<b>P value</b>	0.552	0.878	0.670	0.694	0.083	0.269	0.484	0.687
<b>Haemoglobin</b>								
< 9 gr/dl	80.0±22.7	64.2±38.8	67.8±39.4	51.7±22.4	58.8±20.8	68.7±28.9	67.6±28.3	52.8±25.2
≥ 9 gr/dl	85.4±12.8	56.2±46.6	44.4±47.8	48.7±16.6	60.0±14.7	63.5±26.8	63.9±24.3	55.8±24.2
<b>P value</b>	0.752	0.666	0.174	0.737	0.897	0.515	0.695	0.757
<b>Ferritin</b>								
<1000 ng/ml	92.5±3.5	87.5±17.6	50.0±70.7	52.5±3.5	68.0±22.6	75.0±35.3	72.5±38.8	55.0±28.2
1000-2000 ng/ml	79.7±20.8	57.8±43.3	62.2±40.7	50.2±21.8	60.4±19.6	67.7±29.2	65.0±26.0	51.5±24.7
>2000 ng/ml	89.1±8.0	58.3±40.8	44.4±50.1	50.0±14.8	55.3±9.9	62.5±22.3	68.7±25.5	60.8±23.3
<b>P value</b>	0.339	0.712	0.701	0.955	0.517	0.768	0.891	0.748

PF: physical functioning, PR: physical role, ER: emotional role, VT: Vitality, MH: Mental health, SF: Social functioning, BP: Body Pain, GH: General health

A negative correlation was found between depression and QoL of patients in all subscales (Table 7). A significant difference was found between Beck depression and physical role subscale ( $p=0.042$ ), but the significance was lost, when post-hoc test applied. There was significant difference between the mental health, social functioning and body pain subscales and depression groups ( $p=0.041$ ;  $p=0.014$ ;  $p=0.021$  respectively) but in post-hoc analysis the significance disappeared. Significant difference was found between Beck depression and general health sub-parameter scores ( $p=0.005$ ) and post-hoc analysis showed that the general health score of the minimal

depression group was significantly higher than that of the severe depression group ( $p=0.022$ ). Also, the general health perception of the middle depression group was significantly lower than the minimal depression group ( $p=0.045$ ) (Table 7).

When the correlation between variables was examined, a moderate negative correlation was found between haemoglobin level and emotional role subscale ( $p=0.038$ ;  $\rho=-0.408$ ). No significant correlation was found between SF-36 subscales of other variables (ferritin and BDI) (Table 8).

Table 7. Relationship between quality of life and depression

BDI	SF-36 scales							
	PF	PR	ER	VT	MH	SF	BP	GH
Minimal depression	85.9±13.9	77.2±41.0	75.7±39.6	55.4±20.0	65.0±16.5	77.2±24.8	81.5±17.7	71.8±16.4
Mild	85.6±10.5	68.7±37.2	64.5±38.2	56.8±14.8	66.5±14.1	79.6±17.5	66.8±27.9	48.7±21.3
Moderate	83.0±10.9	35.0±28.5	26.6±43.4	39.0±19.8	54.4±4.5	50.0±12.5	49.5±10.0	41.0±11.9
Severe	63.3±46.1	16.6±28.8	22.2±38.4	33.3±14.4	32.0±21.1	25.0±25.0	37.5±28.8	23.3±20.0
P value	0.716	<b>0.042</b>	0.088	0.186	<b>0.041</b>	<b>0.014</b>	<b>0.021</b>	<b>0.005</b>

PF: physical functioning, PR: physical role, ER: emotional role, VT: Vitality, MH: Mental health, SF: Social functioning, BP: Body Pain, GH: General health

Table 8. Correlation analysis

	Hgb	Ferritin	BDI	PF	PR	ER	VT	MH	SF	BP	GH
<b>Hgb</b>											
rho		0.177	-0.031	-0.060	-0.244	-0.408	-0.109	-0.107	-0.242	-0.265	-0.093
p		0.387	0.880	0.770	0.230	<b>0.038</b>	0.597	0.602	0.234	0.191	0.650
<b>Ferritin</b>											
rho			0.161	-0.033	-0.127	-0.145	-0.088	-0.289	-0.117	-0.032	0.081
p			0.422	0.870	0.529	0.470	0.663	0.144	0.563	0.875	0.689
<b>BDI</b>											
rho				0.197	0.183	0.025	0.204	0.112	0.213	0.028	-0.012
p				0.324	0.361	0.903	0.308	0.577	0.286	0.891	0.953
<b>PF</b>											
rho					0.542	0.455	0.461	0.278	0.539	0.572	0.606
p					<b>0.003</b>	<b>0.017</b>	<b>0.016</b>	0.161	<b>0.004</b>	<b>0.002</b>	<b>0.001</b>
<b>PR</b>											
rho						0.721	0.615	0.463	0.762	0.760	0.679
p						<b>0.000</b>	<b>0.001</b>	<b>0.015</b>	<b>0.000</b>	<b>0.000</b>	<b>0.000</b>
<b>ER</b>											
rho							0.507	0.509	0.708	0.656	0.655
p							<b>0.007</b>	<b>0.007</b>	<b>0.000</b>	<b>0.000</b>	<b>0.000</b>
<b>VT</b>											
rho								0.697	0.586	0.572	0.494
p								<b>0.000</b>	<b>0.001</b>	<b>0.002</b>	<b>0.009</b>
<b>MH</b>											
rho									0.641	0.437	0.378
p									<b>0.000</b>	<b>0.023</b>	0.052
<b>SF</b>											
rho										0.663	0.566
p										<b>0.000</b>	<b>0.002</b>
<b>BP</b>											
rho											0.701
p											<b>0.000</b>

PF: physical functioning, PR: physical role, ER: emotional role, VT: Vitality, MH: Mental health, SF: Social functioning, BP: Body Pain, GH: General health

## DISCUSSION

Periodic screening of patients, their dependence on transfusion, and vascular interventions performed at each visit, mentally exhaust patients with BTM and loss of social time and labour also complicate their adaptation to social life. Phenotypic features and physical difficulties caused by complications can cause them to move away from their social lives. The sociodemographic characteristics (educational status, social status, income status, etc.), physical and mental health status of the patients, play an important role in coping with these processes. As the average age and life expectancy of patients with thalassemia major increases, the concept of quality of life and psychosocial problems become more important (25).

Haghpanah and colleagues had found that, male patients had significantly better QoL than females as in our study; also, high income was a significant factor related to better QoL. Employees were found to have a better QoL on physical role, body pain, mental health and social functioning (respectively  $p=0.011$ ;  $p=0.046$ ;  $p=0.016$ ;  $p=0.037$ ) (26). The QoL of older and married patients was poor in our study, but there was no statistically significant difference. It was observed that the group with higher education level, working in a regular job, with monthly income, without co-morbidity, had better QoL. In short, there was a positive correlation between the academic and personal success rate and QoL, despite the physical limitations of the disease.

In a study investigating depression and QoL in patients with BTM; depression levels were significantly associated with female gender, older age, being single, having higher education, unemployment, and co-morbidity. Mean ferritin levels of the patients were  $1966 \pm 2396$  and there was no relationship between ferritin levels and depression status or quality of life as in our study (27), but, we found that, patients with ferritin value  $<1000$  ng / dl, showed better physical function, physical role, mental health, social functioning and body pain scores. From this point, we can conclude that, the QoL was positively affected if, iron overload was prevented, and adequate chelation was given.

Toret and colleagues had found that, patients with higher education level had better QoL, physical and mental health; also, depression and anxiety symptoms have negatively affected the QoL in Turkish patients (28). We could not find a correlation between educational status and QoL; this could be attributed to low number of patients evaluated in our study.

Safizadeh and colleagues, had found a positive correlation between high salary income and QoL and no correlation between haemoglobin, ferritin levels and QoL (29). We also found no correlation between Ferritin and QoL, but in contrast, a moderate negative correlation was found between haemoglobin levels and emotional role.

Azarkeivan and colleagues showed that, there was a significant negative correlation between depression score

and QoL (30). Similarly, Adib-Hajbaghery and colleagues found a negative correlation between the QoL and depression scores; having low quality of life also caused patients to suffer from stress, anxiety and depression (31). We showed that, the severe depression group had the worst quality of life and there was a significant difference in physical role, mental health, social functionality, pain and general health perception (respectively  $p=0.042$ ;  $p=0.041$ ;  $p=0.014$ ;  $p=0.021$ ;  $p=0.005$ ). Our results seemed to be consistent with the literature.

Oral iron chelation is the most effective therapy that successfully treats transfusion dependent hemosiderosis and prolongs survival in thalassemia patients. Chronic drug usage and background depression could lead to compliance problems. Sobota and colleagues could not find a relationship between compliance with oral chelation therapy and QoL (32). Similarly, no statistically significant difference was found in our patient group ( $p>0.05$ ) but we found that higher depression levels were observed in patients with poor chelation therapy compliance ( $p=0.013$ ). Adib-Hajbaghery and colleagues had also found that depression levels were higher in patients with poor compliance to chelation therapy, so our results were consistent with the literature.

Aydinok and colleagues had found that the rate of depressive disorder of BTM patients was 23 %. Problems such as introversion, depression, anxiety disorders, establishing a social environment in thalassemia patients start at an early age and many of these problems are carried to adulthood. For this reason, individuals should be supported not only physically but also psychologically from the moment of diagnosis (33,34).

Maheri and colleagues showed that, no significant difference in depression scores between male and female sex but, a negative correlation was found between depression scores and education levels. While depression levels of unemployed individuals were higher, depression levels of those who were employed are lower. In addition, depression scores decreased further as patients' position in business life increased (35). We also found that, patients who had a regular monthly income had higher scores in all QoL subscales than unemployed patients. Academic success and economic power seemed to improve the living standards and psychological status of patients, but in contrast to that findings, employed group had significantly higher depression score than unemployed group ( $p=0.044$ ). This could be attributed to lower than expected income of the patients who got a regular job.

## LIMITATIONS

There were many limitations in our study; the number of participants was low and, depression levels were found lower than expected. We have not evaluated whether the patients received psychological support or used antidepressant therapy recently. More significant results might be obtained with studies containing larger number of participants.

## CONCLUSION

In conclusion, patients with severe depression had worst QoL and general health perception of these patients also decreased compared to other groups. High depression scores also significantly related to poor compliance to chelation therapy. Although medical advances in thalassemia treatment have led to an increase in overall survival, patients are still dealing with complications. The physical and mental fatigue experienced by thalassemia patients negatively affect their quality of life and cause psychosocial problems. As with other chronic diseases, increasing the living standards and preventing psychosocial effects in patients with thalassemia should be considered as part of the treatment. For reaching high QoL, better education and economic status, play a role. Better treatment compliance, lower frequency of complications, and lower levels of depression are expected in patients with high QoL. As with many chronic diseases, thalassemia patients should be supported psychosocially and improving their quality of life should be a part of their treatment.

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## REFERENCES

- Martin A, Thompson AA. Thalassemias. *Pediatr Clin North Am* 2013;60:1383-91.
- Williams TN, Weatherall DJ. World distribution, population genetics, and health burden of the hemoglobinopathies. *Cold Spring Harb Perspect Med* 2012;2 a011692.
- Modell B, Darlison M. Global epidemiology of haemoglobin disorders and derived service indicators. *Bull World Health Organ* 2008; 86:480-7.
- Cappellini M-D, Cohen A, Eleftheriou A, et al. Guidelines for the clinical management of thalassaemia: Thalassaemia International Federation, Nicosia (CY); 2008.
- Weatherall DJ. The definition and epidemiology of non-transfusion-dependent thalassemia. *Blood Rev* 2012;26:3-6.
- Kazanci EG, Bektemur G, Bener S, et al. Preimplantation genetic diagnosis in thalassemia and inclusion of the treatment in the scope of reimbursement evaluation. *Health Care* 2017;4:135.
- Tunc B, Cetin H, Gumruk F, Istanbulu B, Yavrucuoglu H, et al. The prevalence and molecular basis of beta-thalassemia in Isparta province and region. *Turk J Pediatr* 2002;44:18-20.
- Origa R.  $\beta$ -Thalassemia. *Genetics in Medicine*. 2017;19:609.
- Langhi Jr D, Ubiali EMA, Marques Jr JFC, et al. Guidelines on Beta-thalassemia major-regular blood transfusion therapy: Associacao Brasileira de Hematologia, Hemoterapia e Terapia Celular: project guidelines: Associacao Medica Brasileira-2016. *Rev Bras Hematol Hemoter* 2016;38:341-5.
- Taher AT, Weatherall DJ, Cappellini MD. Thalassaemia. *The Lancet* 2018; 391:155-67.
- Lucarelli G, Isgrò A, Sodani P, et al. Hematopoietic stem cell transplantation in thalassemia and sickle cell anemia. *Cold Spring Harb Perspect Med* 2012;2: a011825.
- Thalassemia gene therapy nears approval. *Nat Biotechnol* 2019;37:493.
- Belhouli KM, Bakir ML, Saned M-S, et al. Serum ferritin levels and endocrinopathy in medically treated patients with  $\beta$  thalassemia major. *Ann Hematol* 2012;91:1107-14.
- Galanello R, Origa R. Beta-thalassemia. *Orphanet J Rare Dis* 2010;5:11.
- Organization WH. Depression and other common mental disorders: global health estimates. World Health Organization; 2017.
- Beck AT, Steer RA. Internal consistencies of the original and revised Beck Depression Inventory. *J Clin Psychol* 1984;40:1365-7.
- Bilge Y, Kulaksizoglu A. A New Scale: Validity and reliability study of the psychological disorders inventory for adolescents. *J Int Soc Res* 2017;10.
- Hisli-Sahin N. A study on the validity of the Beck Depression Inventory. *Turkish Journal of Psychology* 1988;6:118-26.
- Group W. The World Health Organization quality of life assessment (WHOQOL): position paper from the World Health Organization. *Social science & Med* 1995;41:1403-9.
- Edisan Z, Kadioğlu FG. Health-Related Quality of Life Scales: An Ethical Evaluation. *Turkish Clinics J Medical Ethics-Law and History* 2011;19:8-15.
- Framework IC. The MOS 36-item short-form health survey (SF-36). *Med Care* 1992;30:473-83.
- Kocuyigit H, Aydemir O, Fisek G, et al. Reliability and validity of the Turkish version of Short Form 36 (SF36): Study with a group of patients with rheumatic disease. *Medicine and Treatment* 1999;12:102.6.
- Demiral Y, Ergor G, Unal B, et al. Normative data and discriminative properties of short form 36 (SF-36) in Turkish urban population. *BMC Public Health* 2006;6:247.
- Beck AT, Ward C, Mendelson M, et al. Beck depression inventory (BDI). *Arch Gen Psychiatry* 1961;4:561-71.
- Bush S, Mandel FS, Giardina PJ. Future orientation and life expectations of adolescents and young adults with thalassemia major. *Ann N Y Acad Sci* 1998;850:361-9.
- Haghpanah S, Nasirabadi S, Ghaffarpasand F, et al. Quality of life among Iranian patients with beta-thalassemia major using the SF-36 questionnaire. *Sao Paulo Med J* 2013;131:166-72.

27. Toret E, Karadas NO, Gokce NO, et al. Quality of Life and Depression in Turkish Patients with  $\beta$ -Thalassemia Major: A Cross-Sectional Study. *Hemoglobin* 2018;42:326-9.
28. Gan GG, Hue YL, Sathar J. Factors Affecting Quality of Life in Adult Patients with Thalassaemia Major and Intermedia. *Ann Acad Med Singap* 2016;45:520-3.
29. Safizadeh H, Farahmandinia Z, Nejad SS, et al. Quality of life in patients with thalassemia major and intermedia in kerman-iran (I.R.). *Mediterr J Hematol Infect Dis* 2012;4: e2012058.
30. Azarkeivan A, Hajibeigi B, Alavian SM, et al. Associates of poor physical and mental health-related quality of life in beta thalassemia major/intermedia. *Journal of research in medical sciences: J Res Med Sci* 2009;14:349-55.
31. Adib-Hajbaghery M, Ahmadi M, S P. Health Related Quality of Life, Depression, Anxiety and Stress in Patients with Beta-Thalassemia Major. *Iran J Ped Hematol Oncol* 2015;5:193-205.
32. Sobota A, Yamashita R, Xu Y, et al. 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* 2011;86:92-5.
33. Cakaloz B, Cakaloz I, Polat A, et al. Psychopathology in thalassemia major. *Pediatr Int* 2009;51:825-8.
34. Aydinok Y, Erermis S, Bukusoglu N, et al. Psychosocial implications of thalassemia major. *Pediatr Int* 2005;47:84-9.
35. Maheri A, Sadeghi R, Shojaeizadeh D, et al. Depression, Anxiety, and Perceived Social Support among Adults with Beta-Thalassemia Major: Cross-Sectional Study. *Korean J Fam Med* 2018;39:101-7.