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Health Related Quality of Life in Patients with β-Thalassemia Major in Northern Khuzestan Province, Southwest of Iran in 2015-2016

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ABSTRACT

Introduction: Thalassemia not only influences the patients' physical function, but also their emotional, social, and school functions, leading to the disorder in health-related quality of life (HRQOL) for patients with inappropriate treatment. Therefore, the present study was conducted to assess the health-related quality of life for patients with β-thalassemia major in northern Khuzestan in 2015-2016.

Material and Method: This cross-sectional, descriptive study was conducted in targeted sampling method on qualified patients with thalassemia who recourse the Dezful large hospital thalassemia ward and Andimeshk, Shush, Shoushtar, and Masjid Soleyman (northern Khuzestan) thalassemia centers. Data collecting tools included demographic as well as quality of life SF-36 questionnaires.

Findings: The total score average of quality of life was 63.91 in these patients. A statistically significant relationship was observed between the total average score of quality of life in Persian race, urban patients without hospitalization and/or surgery, compared to others (p<0/05). Pearson correlation test showed a positive, statistically significant correlation between weight and quality of life (r:0/17) (p:0/007).

Conclusion: An appropriate plan for promoting the level of general health as well as training the quality of activity in order for preventing fatigue, along with clinical-remedial services can promote the quality of life.

Key words: Quality of life, Thalassemia major, Khuzestan

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INTRODUCTION

As an autosomal recessive hereditary disorder of blood [1], thalassemia can be identified by synthesis decrease or lack of globin [2,3]. Influenced by globin chain synthesis and also the number and type of engaged chains, clinical signs of this disease are different [4]. Thalassemia major is the most severe form of β-thalassemia which leads to acute anemia [4]. About 60,000 babies with thalassemia will be born around the world every year [5]. Therefore, it is a serious obstacle in general health and highly prevalent in the Mediterranean region, Middle-east, Indian subcontinent, and southeastern Asia [6,7]. Also, it is the most prevalent genetic disease in Iran [8,9]. According to World Health Organization (WHO), about 4% of Iran population are thalassemia gene carriers. Thalassemia dispersal varies in different provinces of Iran. Top 10 prevalent provinces consist of Mazandaran, Gilan, Hormozgan, Khuzestan, Kohgiluyeh and Boyerahmad, Fars, Bushehr, Sistan and Baluchistan, Kerman, and Esfahan [10]. According to statistical data in 2013, there are 3 million transferors and 25,000 patients with thalassemia major in Iran [8]. Based on evaluations, it is more common in Asian developing countries [11]. Thalassemia is a kind of chronic diseases that need blood transition [12]. Iron overload caused by blood transition may make the patient affected severely by the risk of diseases such as heart disease, liver fibrosis [12-14], sexual development insufficiency, osteoporosis, diabetes [12,13]. It's complications on endocrine gland, infections,

short stature, growth, retardation, physical appearance, and premature death affects the disease [11]. Sexual development and fertility either does not exist or is delayed in this disease. Complications such as uncertainty about future and difficulties in long-term planning can be cited as the results of thalassemia major [9,15].

Thalassemia imposes a wide variety of mental and clinical challenges. The aforementioned difficulties not only affect the patients' physical function [13,16] but also their emotional [5], social, and school functions [5,17], leading to disorder in health-related quality of life for those patients that didn't received appropriate treatment [16,18]. It was reported that about 80% of patients with thalassemia suffer from mental problems [3]. Daily treatment with iron chelating and repeated presentation to the hospital is a serious disorder in health-related quality of life by itself [1,18].

According to WHO, quality of life is defined as "individual's perception of their position in life in the context of the culture and value systems in which they live and in relation to goals, expectations, standards, and concerns in life" [9,16]. It has a wide concept that affected physical health, mental position, personal beliefs, social relations, and their interactions with individual's environment [9].

Pakbaz et al. stated that emotional function was one of disorders in quality of life scopes of patients with thalassemia [19]. As mentioned earlier, due to the effects of the disease and its treatment, evidences shows that thalassemia has a negative effect on HRQOL [20]. An evaluation of mental-social health components of HRQOL showed that hemoglobin level before transiting 9 mg/ deciliter or less were associated with lower scores of emotional function [5].

Quality of life disorders in this disease has a negative effect on patients' social life, family relations, job, and activities and increases the risk of hospitalization and deaths from this disease [3]. Observations showed that there was a significant relationship between quality of life and functional level, especially educational function, in these patients [2,21,22]. Shaligram et al. study showed that 44% of patients suffer from mental problems and, 74% of them had a low quality of life [20].

According to Khani et al., 64.9% had no general health and, 20% were suspicious of metal patients. These results showed that patients with thalassemia were at the risk of different psychiatric disorders and, as a result, show the need for psychological consultants [23]. Although some of former studies have investigated the HRQOL issues in patients with thalassemia under usual treatment by blood transition and iron chelating [24], there are a bit contradictory studies about quality of life of patients with thalassemia major, which are mainly on HRQOL of children and teenagers with thalassemia [18]. Since few studies have been done and, due to the necessary evaluation of quality of life level for these patients, researchers have been persuaded to conduct a study for defining HRQOL of patients with β-thalassemia major and its related factors in northern Khuzestan province, southwest of Iran in 2015-2016.

MATERIALS AND METHODS

This study is a cross-sectional descriptive. The study protocol was approved by the Ethics Committee of Dezful University of Medical Sciences and received the Code of Ethics Durs-121. During the study, the researchers were bound to maintain the confidentiality of the secrets of patients and respect the principles of the Helsinki Treaty.

Prior to entering the study, all written consent forms from patients with thalassemia or the parents/guardians of the minors (patients below 16 years of age) who were admitted to thalassemia ward of Dezful large hospital and Andimeshk, Shush, Shushtar, and Masjid Soleyman (northern Khuzestan) thalassemia centers were filled. These patients were selected by targeted sampling method on them with inclusion criteria in the study (aging between 15 years-35 years, literacy, at least one hospitalization.

Data collecting tools consisted of two questionnaires: demographic questionnaire that included age, gender, body mass index, race, educational level, occupation, marital status, monthly income level, hospitalization record, and medication intake and; SF-36 questionnaire, enquiring 36 items, by which the patients' quality of life had been investigated.

The questions of the SF-36 questionnaire include 8 concepts of physical function, social function, role disorder due to physical health, pain, general health, role disorder due to emotional health, energy/fatigue, and emotional well-being. A maximum of 100 and a minimum of 0 is considered for each question. The scores considered for two-option, three-option, five-option, and six-option questions are (50 & 100), (0, 50 & 100), (0, 25, 50, 75 & 100), (0, 20, 40, 60, 80 & 100), respectively. Physical health score is calculated from the mean of physical function, role disorder due to physical health, pain, and general health scopes total scores. Also, the mental health score is calculated from the mean of role disorder due to emotional health, energy/fatigue, emotional well-being, and social function scopes total scores. The mean of total scores of all studied scopes will be calculated to measure the total score of individuals' quality of life. The mean of every scope will be calculated for every individual and, if the mean is lower than 50, the quality of related scope will be considered low and if it is higher than 50, it will be considered high. SF-36 Questionnaire of quality of life is a standard criterion applied in clinical research and practice, evaluation of health care policy, and health status study of general population. Using Cronbach's alpha test, Kafle et al. defined its stability higher than 85% [25]. Montazeri et al. in Iran showed that, except vitality scale (α: 0.65), other Persian type scales of SF-36 had the minimum stability standard coefficients on the range of 0.77-0.9 [26]. Using SPSS-18, independent t-test, ANOVA, and Pearson correlation coefficient, the collected data were analysed.

RESULTS

234 individuals with thalassemia (male: 57/3%, female: 42/7%) with an average age of 23/11 years and the average weight of 51.20 kilogram were enrolled. Other demographics were cited in Table 1.

Table 1: Demographic indices of patients with β-thalassemia major in northern Khuzestan

Variable	Classification	Number	Percentage (%)
Gender	Male	134	57/3
	Female	100	42/7
Marital status	Single	199	85
	Married	35	15
Location	Urban	133	56/8
	Rural	101	43/2
Education	Illiterate	17	7/3
	Less than diploma	106	45/3
	Diploma	71	30/3
	Associate degree	3	1/3
	Bachelor and higher	37	15/8
Hospitalization	Yes	157	67/1
	No	77	32/9
Surgery	Yes	111	47/4
	No	123	52/6
Blood group	A+	80	34/2
	A-	5	2/1
	B+	59	25/2
	B-	5	2/1
	AB+	17	7/3
	AB-	1	0/4
	O+	66	28/2
	O-	1	0/4
Race	Bakhteyari (Lor)	148	63/2
	Arab	21	9
	Fars	41	17/5
	Kurd	4	1/7
	Shushtari	17	7/3
	Ghashghaee Turk	2	0/9
	Other	1	0/4

The total score mean of quality of life was 63/91 for them. Other variables of quality of life and their relations to the gender were cited in Table 2.

No statistically significant relationship was observed between total score of quality of life and gender as well as blood group (p>0/05) (Figure 1).

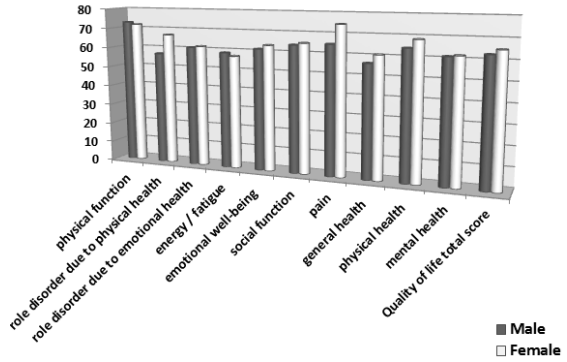

Figure 1: A comparison of mean scores of 8 quality of life scopes related to health, physical health, mental health, and total score of quality of life in patients with β-thalassemia in northern Khuzestan

Table 2: The mean of scores and standard deviation (SD) of 8 quality of life scopes related to health, physical health, mental health, and total score of quality of life in patients with β-thalassemia major in northern Khuzestan and the relationship with gender (Independent T test and Mann-Whitney U were used)

Health dimensions	Mean ± SD	Relation to marital status	Relation to location	Relation to level of education
		p-value*		
Physical function	72/60 ± 26/79	0/76	*0/02	0/48
Role disorder due to physical health	61/32 ± 35/43	*0/08	*0/001	0/67
Role disorder due to emotional health	61/23 ± 35/16	0/41	0/16	0/32
Energy/ fatigue	58/22 ± 24/55	0/08	*0/03	0/22
Emotional well-being	62/28 ± 24/84	*0/01	*0/003	0/18
Social function	64/51 ± 26/35	0/26	*0/003	0/21
Pain	72/65 ± 24/93	0/14	*0/04	0/49
General health	58/47 ±21/37	*0/01	*0/001	*<0/001
Physical health	66/26 ± 20/42	*0/01	*<0/001	0/26
Mental health	61/56 ± 21/96	*0/06	*0/004	0/27
Quality of life total score	63/91 ± 19/45	*0/02	*<0/001	0/32

*Significance level was considered less than 0/05

A statistically significant relationship was observed between quality of life and race, marital status, location, hospitalization, and surgery (p<0/05), because a higher total score mean of quality of life was observed in those urban, Persian race individuals without hospitalization or surgery, compared to others. A statistically significant relationship was observed between general health, emotional well-being, physical and mental health and

marital status (p<0/05). Other relationships were cited in Table 2.

Kolmogorov-Smirnov test showed that the distribution of age, weight, and quality of life variables had been normal (p>0/05). Therefore, Pearson correlation test showed that there was a positive correlation and statistical significance between weight and quality of life (r: 0/17) (p: 0/007). Although there was a positive correlation between age and quality of life (r: 0/01) (p: 0/82), it wasn't statistically significant (Figure 2).

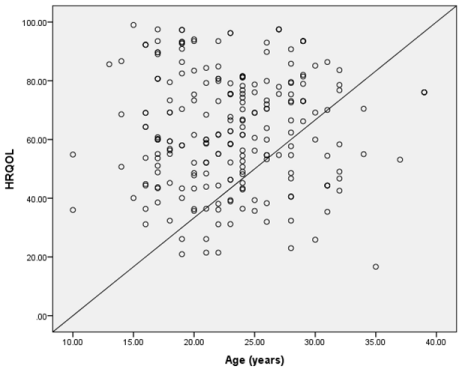

Figure 2: Correlation between age and total score of quality of life in patients with β-thalassemia in northern Khuzestan

DISCUSSION

Even after many years of identifying thalassemia, it continues to be a major health problem around the world, especially in developing and poor countries [27]. Since there are problems such as unpleasant, prolonged, repeated treatment regimens and disease side-effects such as observable changes in craniofacial skeleton in one hand and, increased life expectancy among these patients up to the midlife on the other hand, medical staff face wider mental and social issues in relation to the patients and; patients and their families, in turn, are imposed to various social, emotional and behavioral difficulties [2]. Paying attention to the quality of life, the effect of different psychological considerations, and lifestyle modification can increase the efficiency and independence of individuals to some extent and, help them control numerous disease side-effects and finally treat it [28].

Although the quality of life in patients with thalassemia major is affected by several factors such as family history, family status, and the level of education [3], there was no significant relationship between quality of life score and age, gender, and level of education in the present study. In this regard, Hadi et al. studied HRQOL in patients with thalassemia major and found that, compared to males; female patients took higher scores in social function scope. Also, the greater the age and level of education, the higher will be the scores of physical function and physical role limitation scopes. There was no significant relationship in other scopes [29].

In a study in Thailand, similar to the present one, it was found that quality of life in teenagers with thalassemia was higher than that of young patients [18].

Since supporting the patients with thalassemia requires a national planning, it is recommended to carry out regular screenings for identifying patients, especially those with mental disorders. Also, studying the accessibility of oral chelators of iron is needed [9], because delayed beginning of using iron chelators in childhood decreases the quality of life [9].

Regular monitoring and treating patients' complications, especially heart diseases and hepatitis as well as controlling blood products are very important. Promoting the level of training is effective in improving the quality of life [9]. Despite the increasing developments in screening mental disorders by applying modern treatments which, in turn, improved the quality of life [30], the highest improvements was in social function and energy/fatigue [31].

In the present study the quality of life in the scope of physical health was on an appropriate level, while it is on a low level in many studies [2,32], which persuades governmental policy-makers to pay more attention to the patients and their families and, to give financial and social supports to them.

Quality of life was on an appropriate level in all scopes except general health and energy. These two exceptions reduced total score of quality of life to an average level. Low quality of sleep implies symptoms of depression in patients [33]. This depression is related to physical weakness and mental health of quality of life, however, other diseases and anxiety is related to physical weakness and mental health scope of quality of life, respectively [34].

In the present study there was no significant relationship between quality of life and physical function, while a similar study in Egypt showed that there was a negative correlation between thalassemia and quality of life that, appropriate anaemia control and iron aggregation prevention improved the quality of life [35]. Except role disorder due to physical health, no scope was related to quality of life.

CONCLUSION

The results of this study showed that adults with thalassemia major face problems in general health and energy/fatigue scopes of quality of life. Therefore, suitable plans for promoting general health level and learning how to work in order to fatigue prevention, and clinical- treatment services can improve their quality of life.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the publication of this article.

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