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Beta Talasemi Major Hastalarında SF -36 Anketi Ile Yaşam Kalitesinin Değerlendirilmesi: Tek Merkez Deneyimi

Quality Of Life Assessment By SF-36 Questionnaire In Beta Thalassemia Major Patients: Single Center Study

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Öz

Amaç: Beta talasemi major ağır anemi ile karakterize, yaşam boyu eritrosit transfüzyonu gerektiren kronik bir hastalıktır. Hastalığa bağlı semptomlar, hastalığın komplikasyonları ve uzun süreli tedavinin getirdiği çeşitli zorluklar nedeniyle hastaların yaşam kalitesi olumsuz etkilenmektedir.

Materyal ve Metot: Çalışmaya Aydın Devlet hastanesi Hemoglobinopati tanı ve tedavi ünitesinde izlenen yaşlar 18'in üzerinde talasemi major veya intermedia olan 34 hasta dahil edildi.

Bu hastaların yaşam kalitelerini ölçmek amacıyla Kısa Form-36 (SF-36) Türkçe versiyonu kullanıldı. Ayrıca çalışmaya alınan hastaların demografik ve sosyoekonomik verileri ile yaşam kaliteleri arasındaki ilişkiler araştırıldı.

Bulgular: Çalışmaya yaşları 18-58 arasında değişen 34 hasta dahil edildi. (ortanca yaş 24) Hastaların 26'sı Talasemi major, 8 hasta ise talasemi intermedia idi. Hastaların ortalama aylık transfüzyon ihtiyacı 2,1Ü ve ortalama Ferritin düzeyi 1019 mg/dl(239-6240) idi. 5 hastada (%14,7) diyabet, 6 hastada hipotiroidi (%17,6), 11 hastada (%32,4) hipoparatiroidi, 6 hastada dexa ile bakılan osteopeni (%17,6), 11 hastada (%32,4) osteoporoz, 5 hastada (%14,7) bozulmuş oral glukoz tolerans testi saptandı. Talasemili hastalar sağlıklı Türk populasyonu ile kıyaslandığında sağlığın genel olarak algılanması sosyal fonksiyon, emosyonel sorunlar ve mental sağlığın anlamlı olarak daha kötü olduğu görüldü.

Sonuç: Beta talasemi major hastalarının yaşam kalitesi Türk toplumunun yaşam kalitesine göre daha düşüktür. Kronik hastalıkların varlığı yaşam kalitesini olumsuz etkilemektedir.

Abstract

Objective: Beta thalassemia major is a chronic disorder characterized by severe anemia that requires lifelong erythrocyte transfusion. Disease related symptoms and complications, and the treatment challenges affect the quality of life of the patients.

Materials and Methods: Thirty-four β-thalassemia major or intermedia outpatients, aged 18 and older who admitted to Hemoglobinopathy Diagnosis and Treatment Unit, Aydın State Hospital, Aydın, Turkey were included in this study.

For the assessment of quality of life in these patients, Short Form-36 (SF-36) Turkish version was used. The correlations of the demographic and socio-economic data with the quality of life of the patients enrolled in the study were also investigated.

Results: Thirty-four patients 18-58 years old were included in the study. (Median age was 24 years) Twenty-six were diagnosed with thalassemia major, and eight with thalassemia intermedia. Average monthly amount of transfusion required by these patients was 2.1 units and the mean level of Ferritin was 1019 mg/dl (239-6240). Five patients had diabetes (5; 14.7 %); six had hypothyroid (6; 17.6%), 11 had hypoparathyroidism (11; 32.4%), six had osteopenia as shown by DXA (6; 17.6%), 11 had osteoporosis (11; 32.4%), and five had impaired glucose tolerance (5; 14.7%). Thalassemia patients had significantly poorer scores on perception of health general health, social functioning, and emotional problems and mental health compared to healthy Turkish population.

Conclusion: Individuals with beta-thalassemia major have a decreased quality of life compared to the general Turkish population. Presence of the chronic diseases has negative influences on the quality of life.

Introduction

Beta thalassemia is an autosomal recessively inherited disorder characterized by hemolytic anemia caused by reduced or absence of synthesis of one or more globin chains of the hemoglobine (1).

World Health Organization reported the carrier incidence rate in the world for abnormal hemoglobin as 5.1%. The carrier rate for beta thalassemia is 2.1% for Turkey, located on the region (2,3).

There are three types of the disorder: thalassemia minor, intermedia, and major. Beta thalassemia is the most severe type that who was born with the disorder usually become transfusion-dependent at 6 months or 1 year of age. Beta thalassemia major is a chronic disorder that requires lifelong regular blood transfusion and may cause severe anemia, growth retardation, organomegaly, icterus, skeletal anomalies and a distinct facial appearance. The disorder itself and the related complications necessitates a regular and a challenging treatment regime.

Today, use of intensive erythrocyte transfusion and iron-chelating treatments has improved the life expectancy of thalassemia patients to a good level. The burden of the medical treatments on the patients and the increased life expectancy thus promote the psychiatric problems to develop and/or become manifest. Researchers have reported that among the individuals with thalassemia the rate of psychiatric disorders was 20-80 %.4

Beta thalassemia major is a chronic disorder that can be fatal when untreated. Chronic diseases impair the quality of life of the individual; therefore, the physical, emotional and the social wellbeing.5 While extensive medical treatment has increased the life expectancy it has brought about a call for the improvement of the quality of life in the patients.

Few studies have been done for the assessment of quality of life in the individuals with beta thalassemia. In these studies, the quality of life of the patients was found to be decreased compared to normal individuals (4).

The aim of the present study is to assess the quality of life in beta thalassemia and to contribute to the treatment and the support to the patients with the resultant findings

Short Form -36 (SF-36) is the most commonly used questionnaire for the assessment of quality of life.

Materials And Methods

In this study, 34 outpatients with thalassemia major or intermedia admitted to Adult Hematology Unit, Aydın State Hospital were included. SF-36 was used for measuring quality of life in these patients. SF-36 is a self-assessment scale used for measuring quality of life particularly in individuals with physical disorders. SF-36 explores eight domains of health in 36 items (Table 1).

SF-36 measures the quality of life of the person regarding the last four weeks. It also includes a single item that identifies perceived change in health in the last 12 months. Patients mark their responses on the scale. The application

Table 1. SF-36 Subdomains

	Item number
Physical functioning	10
Physical role limitations	4
Pain	2
Perceived general health	5
Vitality (energy)	4
Social functioning	2
Emotional role limitation	3
Mental health	5

of the scale is easy since the questions are short and clear and can easily be self-administered.

Rating is based on Likert type (3 point/6 point) except for the item 4 and item 5; and on simple yes/no responses for item 4 and item (5). The scale does not give a total score but the scores for 8 subscales are calculated separately. Each subscale is rated between 0-100 scoren (6). The reliability and validity of the Turkish version were made by Koçyiğit et al (7). standardization by Demiral et al (8). Turkish standards are given in Table 2.

Statistical Analyses

Study data were evaluated by Statistical Package for the Social Sciences (SPSS) 11.0 program. Numeric variables with normal distribution were indicated by mean ± standard deviation (SD); numeric variables did not show normal distribution were indicated by median ± interquartile range (IQR).

The influence of categorical parameters on SF-36 subscale scores was evaluated by using Mann-Whitney U (for 2 groups) and Kruskal-Wallis (for more than 2 groups) tests depending on the number of categorical variables. Level of statistical significance was accepted as $p<0.05$ for all assessments.

Results

In this study, 34 outpatients with thalassemia major or intermedia who admitted to Adult Hematology Unit, Aydın State Hospital were included.

Sociodemographic Characteristics

Patients enrolled in the study were consisted of fifteen females (15; 44.1%) and nineteen males (19; 55.9%) (Table 3). The age of the patients was between 18-58 years and the medium age was 24 years.

In educational status classifications, 8 years of compulsory schooling was taken into account. Accordingly the educational status of the patients were classified as primary school (23 patients; 67.6%); high school and equivalents (7 patients; 20.5%); college (1; 3.1%) and illiterate (3; 8.8%) (Table 3).

With respect to the marital statuses the sample were consisted of 30 single (88.2%) and 4 married (11.8%) subjects. (Table 3)

Of the participants, 22 (51%) were student, 12 (27.9%) were employed and 9 were unemployed. Evaluation of the professional lives of the patients showed that the number of

Table 2. SF-36 Turkish Standardizations

	Mean ± SD
Physical functioning	86.6±25.2
Physical role limitations	89.5±29.6
Pain	86.1±20.6
Perceived general health	73.9±17.5
Vitality (energy)	67.0±13.8
Social functioning	94.8±14.2
Emotional role limitations	94.7±20.9
Mental Health	73.5±11.6

Table 3. Distribution of the Sociodemographic Characteristics of the Cases

		N	%
Sex	Female	15	44.1
	Male	19	55.9
Marital Status	Married	4	11.8
	Single	30	88.2
Educational Status	İlliterate	3	8.8
	Primary school	23	67.6
	High school	7	20.5
	College	1	3.1
Live with	Partner-spouse	4	11.8
	Parents	30	88.2
Economic status	0-800 TL	1	3
	800-1200 TL	18	53
	>1200 TL	15	44
Location	City	8	23.6
	Town	6	17.7
	Small town	13	38.3
	Country	7	17.7
Relocation due to treatment requirements?	Yes	6	14
	No	28	86
Employment status	Regular employment	7	17.7
	No regular employment	11	23
	No history of employment	16	35.3
Dismissal due to thalassemia?	Yes	8	35.2
	No	10	29.5
	No history of employment	16	35.3
Total		34	100

persons regularly employed was 7 (17.7%); irregularly employed was 11 (23%); and that 16 patients (35.3%) have no working-occupational history in their life. Table 3 shows the demographic characteristics of the patients.

Thirty patients (88.2%) were living with their parents, four patients (11.8%) with the partner/spouse, and two patients have children. (Table 3).

With respect to total monthly income, one patient (3%) was having an income between 0-800 Turkish Lira, 18 patients (53%) between 800-1200 Turkish Lira, and 15 patients (44%) over 1200 Turkish Lira (Table 3)

All the participants were covered by the social security system and the majority had benefited from social security of either parent. Eight patients (23.6%) were living in a city; six (17.7%) in a town; thirteen (38.3%) in a small town, and seven (17.7%) in rural area. Six patients (14%) previously had to change their place of residence due to their treatment requirements (Table 3)

The amount of monthly transfusion needs was 2.1 units in average, and the mean value for ferritin was 1019 ng/ml. In this group of patients with poor chelation control, the incidence of endocrine diseases was increased. Eleven patients were diagnosed with hypoparathyroidism, six with hypothyroidism, and five with impaired glucose tolerance, five with diabetes, and 11 with osteoporosis measured by DXA (Table 4)

Analyses of the characteristics in the psychiatric histories of the cases revealed that six patients (14%) had a diagnosis of a psychiatric disorder in the past; seven (17.7%) had a history of a psychiatric disorder in a family member. Six patients (14%) had a suicide attempt in their history; six (14%) had a history of self-destructive behavior. Use of alcohol was identified in nine patients (26.7%), and no history of substance use was determined.

Table 4. Co-morbidity assessment of the patients

	No.34
Age -mean (years)	24 (18-58)
Sex	F: 16; M:18
Height -average(cm)	160 ± 10.4
Body weight-average(kg)	54.3 ± 6.8
Splenectomy	24 (70.6%)
Age at splenectomy -mean (years)	10.5 (7-26)
Need for transfusion - average (units)	2.1 ± 0.6
Ferritin (ng/ml)	1019 (239-6240)
Diabetes Mellitus	5 (14.7%)
Hypothyroidism	6 (17.6%)
Hypoparathyroidism	11(32.4%)
Impaired OGTT	5 (14.7%)
Osteoporosis	11(32.4%)

Beta-Thalassemia Major and Treatment Related Features

In the analyses of the distribution of the beta thalassemia and treatment associated characteristics it was found that 14.9% of the patients had transfusion treatment twice weekly, 61.5% in every three weeks and; 23.6% in every four weeks. The average amount of transfusion needs was identified as 2.3 units/month. With respect to the compliance to treatment for the last 6 months, 95.3% of the patients were found to continue to transfusion treatment regularly, and 75% have on regular iron chelation treatment. Two patients (6.2%) have a sibling with thalassemia major.

Chelation treatments received by patients were shown in Table 5.

In the analysis of the scores of the patients on eight subdomains of the SF-36 Quality of life questionnaire, it was found that particularly on the subdomains of general health and emotional role limitations of SF-36 the subjects scored lower than they had on other subdomains. Their scores on the domains of perceived general health, social functioning, emotional problems, and mental health were found to be lower than the scores reflecting normal population (Table 6)

SF 36 parameters were not significantly related to age; sex; splenectomy; ferritin; blood sugar; AST; ALT; creatinine; Ca values; osteoporosis; diabetes; impaired OGTT; use of deferasirox, deferoxamine and deferiprone as chelation treatment; calcium; use of oral contraceptives, antidepressants, testosterone; presence or absence of a psychiatric history; suicidal thoughts; smoking; occupation; level of education, level of education of the parents; place of residence; and income level.

Perceived pain scores were higher in the individuals whose monthly transfusion needs were higher (p: 0.017). Tho-

Table 5. Chelation treatments received by the patients

Chelation treatment	**Number (percent)**
Deferasirox	24 (70.58%)
Deferiprone	1 (2.90%)
Deferoxamine	3 (8.82%)
Deferoxamine- Deferasirox	2 (5.9%)
Deferoxamine - Deferiprone	4 (11.8%)

Table 6.

parameter	Adult Patient (no.34)	Compared to normal population (p value)
Physical functioning	84.6+13.5	0.640
Physical role limitations	84.7+25.1	0.349
Pain	85.2+18.4	0.801
Perceived general health	63.3+25.1	0.006
Energy	71.6+20	0.062
Social functioning	80.7+25.7	0.001
Emotional problems	79.1+28.7	0.001
Mental health	68.6+19.4	0.018
Physiological mean score	51.6+6.7	Turkish mean not available
Mental mean score	51.7+19.6	Turkish mean not available

se with hyperthyroidism showed poorer mental health (p: 0.042), decreased social functioning (p: 0.012); those taking active vitamin D had decreased physical functioning (p: 0.022); digital and diuretic users had poorer physical and social functioning (p values: 0.047 and 0.016 respectively). It was also established that the higher the ejection fraction of the heart, the lower the role limitations due to physical problems (p: 0.041), the higher the energy and social functioning (respective p values are 0.014, 0.020). Perceived pain was found to be higher in thalassemia major patients than in the thalassemia intermedia patients (p: 0.006). Role limitations due to physical problems were found to be higher in those who use alcohol (p: 0.026). Increasing frequency of transfusion was found to be associated with the poorer perception of general health (p: 0.027), and with the decreased level of energy (p: 0.037).

Discussion

Beta thalassemia major is a chronic disorder characterized by severe anemia that requires lifelong erythrocytes transfusion. The disease related symptoms, complications and the challenges of the long-term treatment have a negative impact on the quality of life of the patients with this disorder. Factors such as the time-consuming treatments like blood transfusions and home-based infusion of iron chelation, potential risk of severe complications due to hemosiderosis caused by failing to receive regular chelation treatment, and economic burden of the treatment become highly challenging for the patients.

Quality of life measurement in adult patients with beta thalassemia major were assessed by a limited number of studies (9-11) Unlike our study, other studies usually have selected their samples from pediatric and preadolescent age groups. The sample group in our study mostly composed of adolescents and young adults.

Shaligram et al., in their study, conducted in 2006 with 39 thalassemia major patients 8-16 years old, reported that 74% of the patients had impaired quality of life, measured by European Quality of Life Scale (EQ-5D). They have demonstrated that the identified psychological problems as measured by Childhood Psychopathology Measurement Scale (CPMS) had associated with decreased quality of life (12).

Messina et al., analyzed the SF–36 scores, as in our study, and found no relationship between the scores of SF–36 subdomains and age and sex. They stated that regardless of the age the participants in general scored low on the subdomains of social functioning and role limitations due to emotional problems and that the thalassemia patients had difficulties both in initiating and in maintaining social interactions (13).

In our study, individuals with thalassemia major had significantly lower scores on general health, social functioning, emotional role limitations, and mental health subscales compared to the normal population.

Beta thalassemia major might affect the person's daily activities, familial relations, and professional capabilities due to the challenging nature of the disease itself and of the treatment. Examination of the professional lives of 34 thalassemia major patients in total revealed that only 7 of the 18 patients who had a working life had a regular employment and 16 patients had never had a job. This finding is also consistent with that of the study by Aydın et al. in which the thalassemia patients' need for frequent apply to the hospital for treatment led to decreased performance in social and professional lives.

Jafari et al. have measured the quality of life scores of 200 thalassemia patients with mean age of about 20 years on SF-36 scale and established that majority of patients had decreased total scores regardless of their age and sex (14).

While in the Sobota et al. study, females, elderly, individuals having too many complications due to the disorder, and those with many side effects due to iron chelation had lower quality of life scores. In their study, they found significantly lower scores on five subdomains of the 8 domains (physical functioning, physical role limitation, general health, social functioning, and emotional role limitation), and general physical and mental health (15).

Yet, in another study, which included 179 beta thalassemia (major and intermedia) patients, SF-36 scale was administered and through multiple regression analysis, it was demonstrated that the somatic co-morbidities and scores on depression scale were associated with the poorer physical quality of life; and scores on depression and anxiety scales were associated with the poorer mental quality of life (16).

In our study, consistent with the Jafari et al. study, age and sex had no influence on quality of life.

Perceived pain scores were higher in the individuals whose monthly transfusion needs were higher (p: 0.017). Those with hyperthyroidism showed poorer mental health (p: 0.042), decreased social functioning (p: 0.012); those taking active vitamin D had decreased physical functioning (p: 0.022); digital and diuretic users had poorer physical and social functioning (p values: 0.047 and 0.016 respectively). It was also established that the higher the ejection fraction of the heart, the lower the role limitations due to physical problems (p: 0.041), the higher the energy and social functioning (respective p values are 0.014, 0.020). Perceived pain was found to be higher in thalassemia major patients than in the thalassemia intermedia patients (p: 0.006). Role limitations due to physical problems were found to be higher in those who use alcohol (p: 0.026). Increasing frequency of transfusion was found to be associated with the poorer perception of general health (p: 0.027), and with the decreased level of energy (p: 0.037).

In our study, no correlation between the scores on subdomains of SF-36 and the level of ferritin, and the type of iron-chelating agent and monthly income. This result is contrasted with the results of the Dahlui et al. study. In their study, transfusion-dependent thalassemia patients who were on optimum dose of deferoxamine had longer 'quality-adjusted life years' (QALYs) than the patients who were on sub-optimum dose of deferoxamine, and reported that QALYs were associated with serum ferritin, complications due to iron loading and total household income (17). In our study, presence and absence of a psychiatric history, presence of suicidal thoughts, use of antidepressants, smoking, occupation, level of education, parental level of education, place of residence, and monthly income did not show any significant differences with respect to the parameters of SF-36.

Outpatients followed with the diagnosis of beta thalassemia major can be administered SF-36 scale and their treatment can be arranged accordingly, and can be referred to Psychiatry services when considered necessary.

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