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Research Article

Quality of life among Ardabil patients with beta-thalassemia major

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ABSTRACT

Background: Thalassemia as the most common genetic disorder worldwide is regarded as a serious problem in public health issues in the Mediterranean region. Patients with beta-thalassemia major experience physical, psychological and social problems that lead to decreased quality of life. The aim of this study was to assess health-related quality of life and its determinants among patients with major beta-thalassemia.
Methods: This was a population-based cross-sectional survey of quality of life. Population with thalassemia major (aged ≥ 2 years) of both genders who had records in Thalassemia Clinic of Bu-Ali Hospital, and those who regularly refer for blood transfusion or follow-up visits. Data were collected from December 2013 to May 2014.The self-administered short form-36 (SF-36) questionnaire was used to measure quality of life in patients with thalassemia. Data were analysed using descriptive statistical tests (mean, SD, and frequency), and inferential statistical test (t-test) in SPSS.17 software.
Results: Our samples were 20 men and 23 women. The median age was 20 years (2-42). After reviewing the patients' quality of life, it was observed that the mean score for physical function was 79.8, role limitations due to physical reasons 78.8, bodily pain 74.4, general health 59.1, fatigue or vitality 63.3, social function 70.21, role limitations due to psychological reasons (emotional) 77.3 and mental health 65.4. On two scales, role physical (P = 0.33) and role emotional (P = 0.13), the men showed significantly lower scores than the women.
Conclusions: After reviewing the patients' quality of life, the highest quality in physical function and lowest quality in general health of patients were observed. In the quality of care data all scales were in very good level except general health.

Keywords: Iran, Thalassemia major, Quality of life, Short form-36

INTRODUCTION

Thalassemia as the most common genetic disorder worldwide is regarded as a serious problem in public health issues in the Mediterranean region, Southeast Asia, the Indian subcontinent and the Middle East, including Iran.[1,2] Iran is located on the thalassemia belt in the world and thalassemia carriers are evaluated from 1 to 10 percent [average % 4.5] transitively.[3] According to the statistics, the number of patients with thalassemia major (the most severe form of thalassemia) in Iran is more than 20,000 individuals.[4] Thalassemia affects the synthesis of globin chain and according to the number and type of the involved chains, clinical symptoms vary. Beta-thalassemia is the most common frequent type of thalassemia which presents in three forms: thalassemia minor which leads to a mild asymptomatic haemolytic anaemia, thalassemia intermediate and major.

Interestingly, thalassemia major is the most severe form of beta-thalassemia leading to severe anaemia and patients are in need of blood transfusion since the young age; this type may end in heart failure , due to iron overload, or early death in childhood in case of no transfusion.[5] Nowadays, with improvements in the treatment of patients with thalassemia, these patients have a longer life expectancies and a larger number of them reach older ages.[6] As a result of this increase in life expectancy, their needs also change and entities such as continuing education, career and family making become more prominent amongst them.[7] It is then anticipated that effective and suitable life background would be prepared for them in the society and they become as active as other members of the society.[8] An increase in the life expectancy is accompanied by certain challenges such as bone diseases, infertility, consecutive referrals for blood transfusion, subcutaneous infusion of chelators, and moral stresses, which these aforementioned problems would affect mental, physical, social and educational functions of these patients.[1,7] Hence, authorities who provide services to patients should be aware of the related mental and social consequences in addition to the burden of this disease, just like any other chronic disease, to prepare better living environment for these patients.[8] Therefore, in this study, we aimed to measure the Quality of life in patients with beta-thalassemia major using SF-36 for the first time in who referred to Thalassemia Clinic of BU-Ali Hospital in Ardabil, Iran.

METHODS

This is a cross-sectional study that has been done on patients (aged ≥ 2 years) with major beta-thalassemia of both genders attending the Haematology & Oncology Center of Bu-Ali Hospital in Ardabil province, located on the North West of Iran. Data were collected between December 2013 and May 2014. The total sample was 43. No sample size calculations were carried out prior to the study. We assessed Thalassemia Major Patients who were dependent on blood transfusions, and who visited for monthly blood transfusions and clinical examinations. Thalassemia major diagnosis was based on the results of electrophoresis of blood haemoglobin. The study protocol was approved by both the Institutional Review Board and the Ethics Committee of Ardabil University of Medical Science. Informed written consent was obtained from all patients. For data collection, questionnaires were used. The researcher-made demographic questionnaire included age, gender, educational level employment status, marital status. The Persian version of the SF-36 questionnaire was used in this study, and this had previously been translated from English and validated (9).This questionnaire evaluated the general health of thalassemia major patients according to 8 categories: physical functioning (PF), role-physical (RP), bodily pain (BP), general health (GH), vitality (VT), social functioning (SF), role-emotional (RE) and mental health (MH). The Cronbach alpha for the reliability of this questionnaire has been found to range from 0.77 to 0.90 in Iran and from 0.65 to 0.96 in other countries.[10] The total score of these 8 indices ranged between 0 and 100, with designations of weak (≤20), bad (21-40), good (41-60), very good (61-80), and excellent (>81).[10] Questionnaires were written through interviews and completed by trained students. Explanations about the disease and reasons for conducting this study were given to the patients, and permission to complete the questionnaire was obtained from the patient and parents. Literate patients were asked to complete the questionnaire themselves if they wished. Those who were illiterate were assisted by a trained researcher. Considering the young age of the patients, questions were asked, and interviews were conducted in such a way so as not to undermine integrity and independence of questions. Data was analysed using descriptive statistics (frequency, mean, and standard deviation) and inferential statistics (t-test) in SPSS-16 software. P-value of less than 0.05 was considered statistically significant.

RESULTS

Table 1: Socio-demographic data of samples.

Age (years)	n	%
2-12	5	11.6
12-2	19	44.1
22-32	16	37.2
≥ 32	3	6.9
Gender		
Male	20	46.5
Female	23	53.4
Marital status		
Single	25	58.1
Married	18	41.9
Level of education		
Illiterate	3	7
Primary	26	60.5
Intermediate	10	23.2
Secondary	4	9.3
Employment status		
Student	8	18.7
Retired	0	0
Employed	4	9.3
Jobless	31	72
Economic class		
Low Income	12	27.9
Moderate Income	9	20.9
High Income	22	51.2

In our study, we included 43 patients with beta-thalassemia major aged between 2 and 42 years of age (mean =20.32 and SD =8.47) (range, 2– 42 years). Males made up 46.5% of our sample. 25 subjects (58.1%) were single and 18 (41.9%) was married. Table1 showed demographic characteristics of participants. The mean

scores on the 8 indexes of the SF-36 Test are shown in Table 2. Physical functioning index had the highest mean score [79.8 (SD 31)], while General Health had the lowest mean score [59.1 (SD 20.7)]. Scores between 80 and 100 for each of the 8 indexes were achieved by 27 patients (62.7.9%) for physical functioning, 28 (65.1%) for Role-physical, 19 (44.1%) for bodily pains, 9 (21%) for general health, 10 (23.2%) for vitality, 15 (34.8%) for social functioning, 27 (62.7%) for role-emotional, and 14 (32.5%) for mental health. According to the analysis, although the female participants had a higher score in all domains except for RE & RP, when compared to the male participants, such a difference was only significant in SF and GH domains ($p<0.05$)(Figure 1). Unlike income, there wasn't any significant relationship between age and the quality of life scores ($p>0.005$).

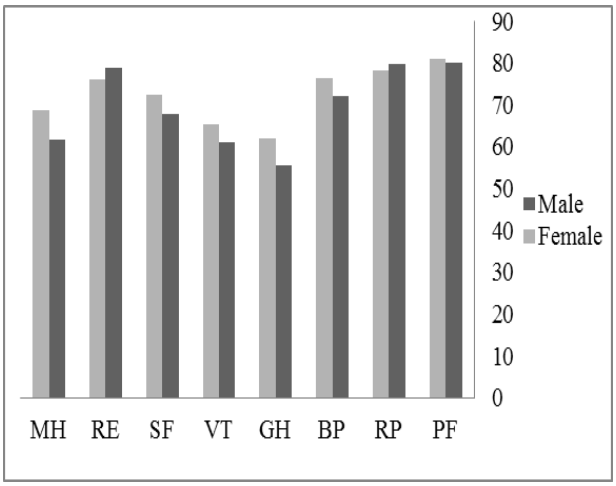

Figure 1: SF-36 domain scores in male and female patients.

DISCUSSION

Thalassemia Major has become a major general health problem throughout the world, especially in developing and poor countries.[9] Thalassemia major is usually recognized in childhood, at which time patients begin treatment. Over their lives, however, patients and their families are at risk for social and behavioural disorders. Due to distress from both the illness itself and the need for iron chelation, thalassemia subjects frequently display maladaptive coping strategies and high levels of anxiety with psychosocial dysfunction.[10] The quality of life amongst the patients with thalassemia in Ardabil is very good in eight domains under study except general health compared to the rest of Iranian healthy members (Table 2). Such a difference is not surprising since due to special physical conditions and problems related to their disease, patients are not anticipated to have a very good quality of life.[8] The results of a study conducted by Safizadeh et al, on the quality of life among patients affected with thalassemia major aged sixteen years and older, in Kerman city revealed that they had a good quality of life in all domains, as in his study the highest score was seen in PF and SF, and the lowest score in BP and MH.[11] Another study conducted by AzarKeivan on the quality of life among patients affected with thalassemia (major and intermediate) aged eighteen years and older, in Tehran city revealed that they had a very good quality of life in all domains except for MH and BP compared to this present study, in her study the highest score was seen in PF and SF, and the lowest score in BP and MH.[12] In a recent report by Hadi et al, it was reported that in Shiraz has demonstrated a better quality of life in all domains compared to this study, the highest score was seen in RP and Pf and the lowest score in GH and V.[13] The only common point between these three studies and the present study was in PF domain which was better than the other domains; hence it seems that receiving suitable treatments would decrease the patients' physical problems.

Table 2: Health-related quality of life of the patients with beta-thalassemia major, according to the Short Form-36 (SF-36) score.

Score	Total (n = 43)	Men (n = 20)	Women (n = 23)	P-value
Physical Functioning (PF)	79.8 ± 31	80.1 ± 33.1	81.1 ± 28.1	0.25
Role-Physical (RP)	78.8 ± 18.5	79.7 ± 14.7	78.1 ± 21.6	0.33
Bodily Pain (BP)	74 .4 ± 22	72.1 ± 21.9	76.4 ± 22.4	0.49
General Health (GH)	59.1 ± 20.7	55.7 ± 22.1	62.1 ± 19.4	0.03
Vitality (VT)	63.3 ± 20.1	61.2 ± 20.2	65.2 ± 20.3	0.17
Social Functioning (SF)	70.21 ± 25.9	67.7 ± 27.5	72.3 ± 24.9	0.01
Role-Emotional (RE)	77.3 ± 27.7	78.7 ± 26	76.1 ± 29.6	0.13
Mental Health (MH)	65.4 ± 19.1	61.6 ± 23.3	68.8 ± 14.2	0.37

In the present study, mean physical function (PF) score was 79.8, which is higher than the 69.1 reported by Ismail et al, who assessed the health-related quality of life in Malaysian children with thalassemia.[14] Similarly, Caocci et al, reported a PF of 68.4 in a cohort of thalassemia patients from Syria, Palestine, and Iraq.[15] In another study by Gharaibeh et al, a PF score of 54.2 in a cohort of Jordanian children was observed.[18] Also in a study conducted by Amoudi et al, in Western Saudi

Arabia PF score was 61.4.[16] However, all of the above findings disagrees with the results obtained in this study.PF score in Studies conducted in Iran by Khani et al, in southern coast of the Caspian sea was 73.58. Another study by Safizadeh in Kerman on the 209 patients with thalassemia major, was 72.11 and in southern Iran on the One hundred and one patients with beta-thalassemia major was 86.9.[11,17,18] The pattern in the RP score comes in tandem with PF score in our sample and in study conducted by Hadi et al, our sample concluded an RP score of 78.8, compared to 54.31 in the Southeast Iran study (n=13), 65.4 in Southern Iran(n=21), 59.06 in Northern (n=20) & 74.2 in the Italian study (n=24).[13] According to our findings of general health scale compared with other scale the lowest score to Assigned to the same view of the findings of Gollo in Italy in 2001 and 2009.[19] In our study, higher income was a significant determinant of higher quality of life scores for the physical and mental health dimensions, as well as higher total scores. This was concordant with the results from Tajvar et al.[20] In a study on an elderly Iranian population. Obviously, with better economic status, different populations and especially thalassemia patients would be able to confront their disease-related problems more easily. Among the population studied, 55.8% were not more than 22 years old. We did not find any significant correlation between age and the quality of life scores. Sobotaet al Investigated quality of life in a longitudinal thalassemia cohort aged over 14 years, compared with the norms in the United States, along with the influence of clinical factors.[21] Differing from our results, they showed worse quality of life among older patients. The results have also demonstrated that sex is influential in GH domain and the girls' status seemed to be better. Although it sounds that problems resulting from thalassemia and its treatment are not any different between girls and boys, the expectations from the males in the society (such as managing a family, income, etc.) in Iran can justify these differences. In a study by Hadi, women had a better quality of life in SF domain whereas studies conducted in Thailand and Malaysia has shown that sex is not effective on the quality of life domains.[1,13,14]

CONCLUSION

All patients with thalassemia should undergo assessment of quality of life amongst so that interventions focusing on the affected domains can be implemented. In addition, patients with thalassemia and their families often require support in many aspects, especially psychological support to prevent mental disorders.

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