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Evaluation of The Quality of Life of Patients With B-Thalassemia Major Referring to Mofid Children's Hospital In 2010-2011

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Abstract: Objective: Determining degree of health impairment as perceived by the β-thalassemia major patients is essential information needed to recommend suitable therapy. Due to limited research in this area, this study was done and the aim was survey of quality of life in patients with β-thalassemia major.
Materials and Methods: In this cross-sectional study, 70 β-thalassemia major patients aged 15 years and over referred to Mofid children hospital of Tehran in 2010-2011 were studied using a demographic and Global Assessment Scale and Iranian version of SF-36 questionnaires. The samples were chosen on the basis of an available non-randomized sampling. The data was analyzed using SPSS software and statistical analysis methods.
Results: The mean age of 70 subjects enrolled in this study was 20.0 (SD=4.0) years. 42 patients (60%) were male and 28 patients (40%) were female. The analysis showed that there was no significant association between the gender and age groups and age at the first blood transfusion and the presence of co-morbidity with quality of life and Global Assessment Scale. All of the patients acquired scores above 70 in the Global Assessment Scale. Quality of life of patients was low in the Physical Function and Bodily Pain scales of Physical Health component in comparison with healthy individuals but patients had favorable quality of life in the Mental Health component.
Conclusion: Presented data suggested that for improvement of quality of life in β-thalassemia major patients, special attention regarding physical aspects and better accomplishment medical and rehabilitation services is necessary in addition to psychological problems of these patients.

Key words: β-thalassemia major, quality of life, the SF-36 questionnaire, Global Assessment Scale

INTRODUCTION

Thalassemia is one of the problems of our modern society and many countries in the world. Thalassemia is a hereditary anemia that, due to interruption in the production of globin chains in the hemoglobin building, reduces the life span of red blood cells. The most common form of thalassemia in Iran is β-thalassemia, and its

severe form is ß-thalassemia major, also called Cooley's anemia. Regarding the frequency of thalassemia gene and the presence of 20,000 patients with ß-thalassemia major in Iran and coverage of 160 patients in a thalassemia clinic in Mofid children's hospital in Tehran, the quality of life of these patients is important in providing care services. Because of severe anemia, these patients require frequent blood transfusions to survive, and blood transfusion-induced hemosiderosis requires the continued use of iron chelators, and other complications of the disease, such as infection and endocrine disorders, and impaired fertility and heart failure require careful medical care [1-5]. In spite of many advances in the field of recognizing and examining the psychological problems of chronic diseases, limited studies have been done in identifying psychological disorders and assessing the quality of life of patients with ß-thalassemia major [6-10]. By increasing the life expectancy of these patients due to the improvement of the therapeutic process, paying attention to psychological problems and assessing the quality of life of these patients has become an important issue. The chronic course of this disease requires the normal living conditions in various fields, including social activities, having a proper job position, marriage and family formation, and lack of proper social support can be a factor in the emergence of a variety of psychological problems such as anxiety, depression, and other consequences, which certainly require the attention and follow up of the issues [6-10]. In chronic diseases like thalassemia major, in addition to controlling the symptoms of the disease, improving the quality of life is very important. The effect of the physical and emotional pain of the disease on the individual is not entirely recognizable from the viewpoint of the physician and the nurse, and depends on the patient's own true feelings, and not enough attention can be given to the different aspects of the disease for patients and their families [11]. Evaluation of the quality of life of patients with ß-thalassemia major is a relatively new issue and today, decreasing the mortality rate of these patients as a result of the progress of medical treatment is an important indicator in the provision of health care. Therefore, considering the importance of the above issues, the present study was conducted with the aim of evaluating the quality of life and performance of patients with ß-thalassemia major. The results of this study can lead to improved health services and rehabilitation and improve the quality of life of these patients. These patients can achieve the desired mental health and well-being, by having more favorable social situation and ultimately living experience similar to those of normal population standards.

Materials and methods

All patients with ß-thalassemia major aged 15 years and over who were referred to the Mofid Children's Hospital, the sample size was calculated based on the following formula and equal to 67 people. All patients with ß-thalassemia major dependent on blood transfusion of 15 years old and above who were referred to Mofid Children's Hospital after receiving written consent were included in this study. The variables included age, sex, age of onset of the first blood transfusion, hepatitis C, B infection and HIV infection, heart failure and endocrine disorders including diabetes, hypothyroidism and hypoparathyroidism were recorded by interview, examination and information turned out in patients' records in the information form. Patients' quality of life was determined based on SF-36 questionnaire (translation and validation of Persian version) [12] and general performance evaluation scale [13]. The composition of the SF-36 questionnaire is that the questions and concepts of the ground, the structure of the scale, and the measurements of the summary at three levels are classified as follows: 36 total questions, which are in 11 major questions.

Eight indicators and two measures of the survey include:

1. Physical health: Physical performance, role limitation due to physical problems, the amount of physical pain, general health

2. Mental health: The rate of social performance, the amount of cheerfulness and vitality, Limitations of role due to mental problems, mental health

In the SF-36 test, each question only applies to the scoring of a scoring. For some questions, scores are re-encoded so that all scales get one-way scores. The scores for each scale range from zero to 100, that zero indicates the worst and 100 reports the best for the desired scale. Also its reliability and validity were confirmed.

Global Assessment of Functioning (GAF) and Children Global Assessment of Scale (CGAS) are general performance evaluation measures that are used in psychiatry to evaluate psychosocial function. The GAF was designed in the early 1990s which is related to Axis V of DSM-IV and provides a measure of the overall performance of psychiatric symptoms. The GAF is very similar to GAS, which is used for the same purpose in DSM-III -R and is based on the severity of symptoms and social function and job performance, and in fact examines the psychological, social, and occupational performance of the individual [3,13]. A sample of the SF-36 questionnaire, as well as a general scale of performance are provided in the appendices. The data were analyzed using SPSS statistical software in descriptive and analytical statistics.

Statistical methods and statistical tests

In this study, quantitative variables have been used for mean and standard deviation, and for qualitative variables, frequency and proportion have been used. In order to study the normal values of quantitative variables, the Kolmogorov-Smirnov Z test, the relationship between variables and the comparison of the means by Mann-Whitney Test, t-test and the comparison of the differences between groups were used by χ^2, Fisher's exact test and Pearson correlation. P-value <0.05 was considered significant.

Results

The aim of this study was to evaluate the quality of life and function of patients with ß-thalassemia major on 70 patients aged 15 years and over who were referred to Mofid Hospital between years 2019-2011. 42 (60%) were men and 28 (40%) were female (Figure 1).

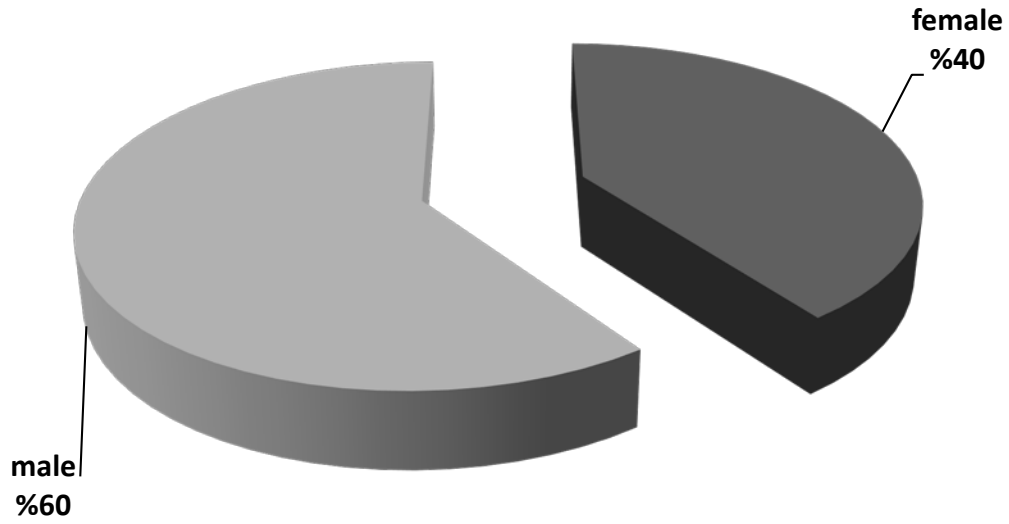

Chart (1): Distribution of Sexual Prevalence among Thalassemia Major Patients Referred to Mofid Hospital in 2010-2011

The mean age of the patients was 20 ± 4 years with a range of 15-31 years (Table 1 and Chart 2). The mean age of the patients was 27.9 ± 29 months with a range of 6 to 120 months in the first transfusion (Table 1).

Table 1: Mean, standard deviation and age range of patients with thalassemia major and age of first blood transfusion referring to Mofid Hospital during 2010-2011

Variables	*Average*	*Standard deviation*	*Domain*
Age (year)	20/0	4/0	15-31
Age of first blood transfusion (month)	27/9	29/0	6-120

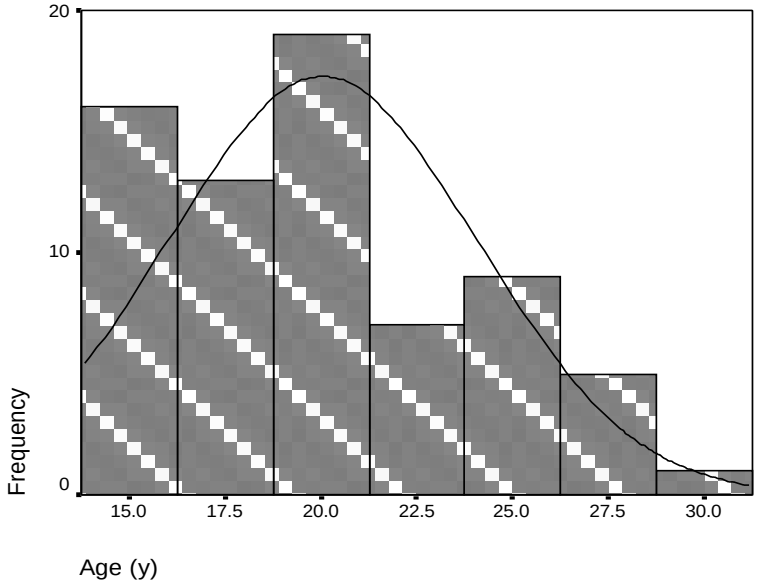

Chart No.2: Age distribution of patients with thalassemia major referring to Mofid Hospital in 2010-2011

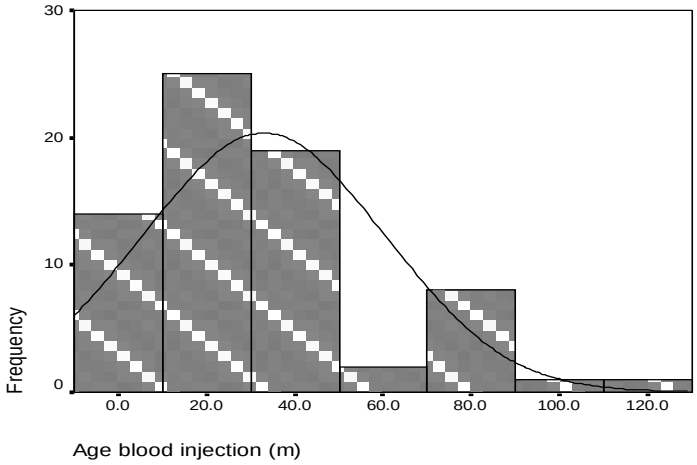

Diagram (3): Age distribution of patients with thalassemia major during the first blood transfusion referring to Mofid Hospital in 2010-2011

The results of the related illnesses in patients with ß-thalassemia major in Mofid Hospital were as follows: 7 (10%) Hepatitis C, 1 person (1.4%) Hepatitis B, 6 (8.6%), diabetic, 5 (1 / 7%) hypothyroidism, 2 (2.9%) hypoparathyroidism, one person (1.4%) had heart failure, and no cases of HIV infection (Figure 4).

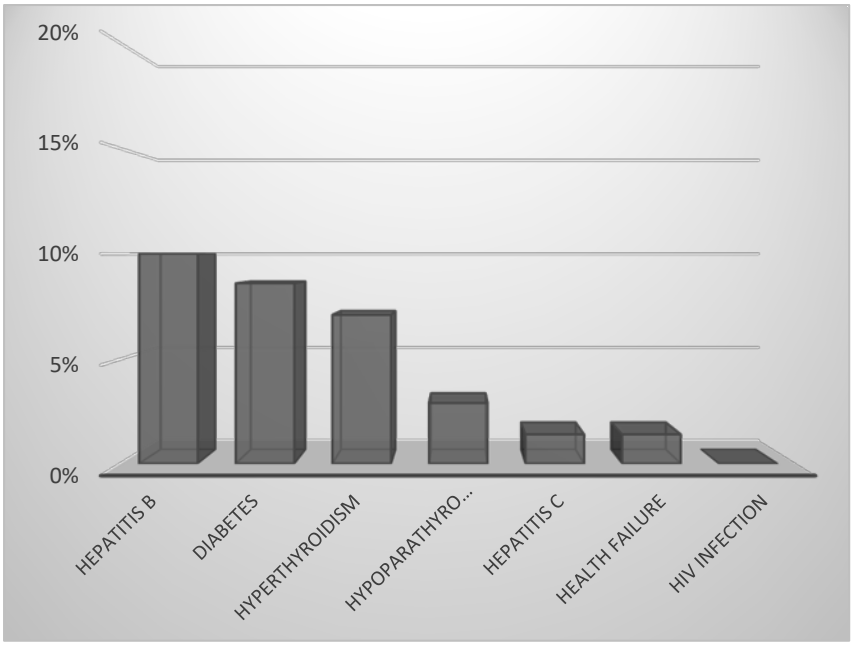

Diagram (4): Distribution of the prevalence of associated diseases in patients with thalassemia major referring to Mofid Hospital in 2010-2011

On the overall scale, 39 people (56%) scored a score of 91-100, meaning superior performance in all domains, at home, at school, or with peers. 29 people (41%) scored 81-90%, meaning good performance in all domains and with probable transient and controllable problems such as mild anxiety. Two people (3%) earned a score of 71-80, meaning mild degradation at home, at school, or with their counterparts. None of the people surveyed earned a score of less than 70 (Figure 5).

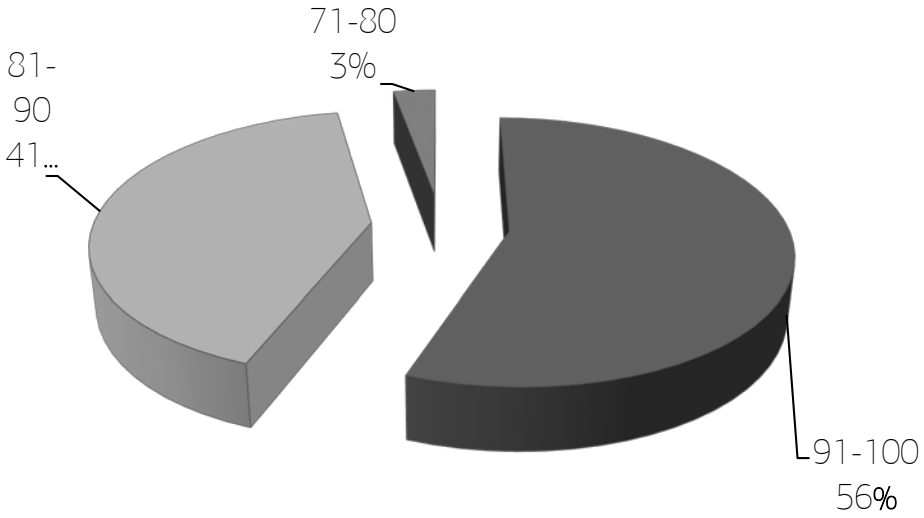

Diagram (5): Distribution of Frequency Percentage of General Scale of Function in Patients with Thalassemia Major Referred to Mofid Hospital in 2010-2011

The Cronbach's alpha coefficient, which indicates the reliability of the internal consistency of the question about each measurement, was found to be greater than the recommended coefficient of 0.7 (in the case of translation of the SF = 36 test in Iran), which shows that, there is a relationship between limitation of role due to physical problems with physical. Function [12]. In this study, the Cronbach's alpha coefficient was equal to 0.799. There was a relationship between different scales: there was a relation between general health and physical pain. There was a relationship between happiness and vitality with physical and general health. There was a relationship between social performance and vitality. There was a relationship between role limitation due to mental problems and vitality and social function. Mental health was related to the following: physical pain, general health, well-being, social function, and role limitation due to mental problems.

In Table 2, internal scale correlation coefficients for measuring the standard tool SF-36 is shown.

Table 2: Distribution of internal correlation coefficients of different dimensions of SF-36 questionnaire in patients with thalassemia major referring to Mofid Hospital during 2010-2011

Dimensions of the questionnaire SF-36	*Physical function*	*Physical problems*	*Physical pains*	*General health*	*Social performance*	*Joy and delight*	*Mental problems*	*Mental health*
Physical function	1/0							
Physical problems	0/46	1/0						
Physical pains	0/15	0/15	1/0					
general health	0/13	0/22	0/42	1/0				
Joy and delight	0/21	0/20	0/45	0/59	1/0			
Social performance	0/45	0/35	0/19	0/17	0/36	1/0		
Mental problems	0/27	0/60	0/18	0/18	0/36	0/42	1/0	
mental health	0/33	0/15	0/42	0/44	0/71	0/44	0/32	1/0

The results of descriptive statistics of different dimensions of SF-36 questionnaire in β-thalassemia major patients referring to Mofid Children's Hospital are summarized in Table 3. As can be seen, the highest percentages are related to the average physical and social performance, and the lowest percentages were related to the average overall health and vitality.

Table 3: Distribution of internal correlation coefficients of different dimensions of SF-36 questionnaire in patients with thalassemia major referring to Mofid Hospital during 2010-2011

Dimensions of the questionnaire SF-36	*Average*	*Standard deviation*	*minimum*	*Maximum*
Physical function	77/6	25/9	0/0	100/0
Physical problems	68/2	33/5	0/0	100/0
Physical pains	71/4	18/6	20/0	100/0
general health	65/9	19/3	10/0	100/0
Joy and joy	66/4	20/2	15/0	100/0
Social performance	77/7	18/4	12/5	100/0
Mental problems	70/0	35/5	0/0	100/0
mental health	70/9	19/8	16/0	100/0

The results of comparing the scores of quality of life dimensions of patients with β-thalassemia major were evaluated using SF-36 questionnaire with percentages of healthy subjects (4163 healthy individuals studied by Montazeri) in Table 4. The mean of quality of life scores in patients with β-thalassemia major was lower in physical activity and physical pain than in healthy subjects. There was a significant difference between the mean scores of these two dimensions among healthy subjects with β-thalassemia major. However, there was no significant difference between the two groups regarding the other dimensions.

Table 4: Comparison of mean percentage of scores of different dimensions of SF-36 questionnaire in patients with thalassemia major referring to Mofid Hospital during the years 2010-2010 with healthy people

Group of people Dimensions of the SF-36 questionnaire	*healthy people people 4163)*	*Patients with Thalassemia Major (70 people)*	*Significance level*
Physical function	85/20±3/8	77/25±6/9	0/003
Physical problems	70/38±0/0	68/33±2/5	0/694
Physical pains	79/25±4/1	71/18±4/6	0/009
general health	67/20±5/4	65/19±9/3	0/515
Joy and joy	65/17±8/3	66/20±4/2	0/633

Social performance	76/24±0/4	77/18±7/4	0/774
Mental problems	65/41±6/4	70/35±0/5	0/377
mental health	67/18±0/2	70/19±9/8	0/073

The mean scores of different dimensions of quality of life based on SF-36 questionnaire of patients with β-thalassemia major were compared in two groups of women and men. There was no significant difference between the mean scores of different aspects of quality of life according to the SF-36 questionnaire and the gender of patients with β-thalassemia major (Table 5).

Table 5: Comparison of mean percentage of scores different dimensions of quality of life based on SF-36 questionnaire by gender segregation of patients with thalassemia major referring to Mofid Hospital in 2010-2011

The gender of the patients *Dimensions of the SF-36 questionnaire*	*Men* *standard deviation) ±)*	*Women* *standard deviation) ±)*	*significance level*
Physical function	79/26±0/1	76/26±5/0	0/666
Physical problems	67/34±5/2	68/33±7/4	0/886
Physical pains	70/18±3/3	72/19±2/0	0/617
general health	61/19±3/5	69/18±3/4	0/079
Joy and joy	69/18±5/1	64/21±1/6	0/274
Social performance	77/15±1/8	78/20±1/4	0/513
Mental problems	72/34±2/0	68/36±3/9	0/744
mental health	71/19±5/2	70/20±5/5	0/842

There was no significant difference in the overall performance scale (GAS) among patients with β-Thalassemia major referring to Mofid Hospital between men and women (P = 0.778) (Figure 6).

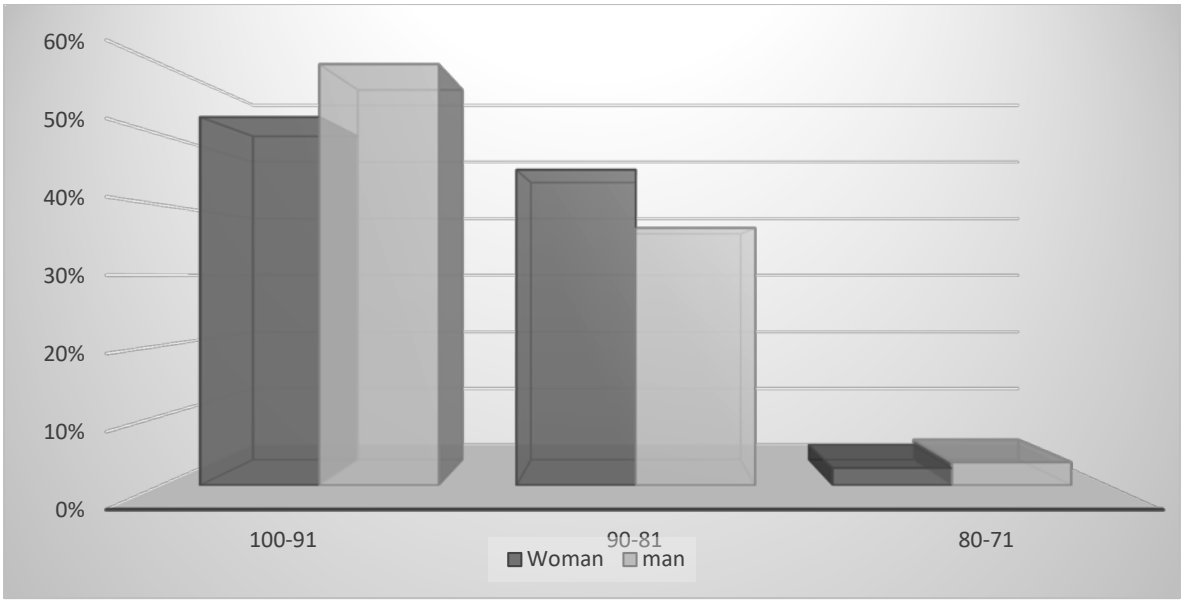

Chart (6): Comparison of the frequency percentage of general measures of functional performance by gender in patients with thalassemia major referring to Mofid Hospital during 2010-2011

Comparison of the mean percentage of different dimensions of quality of life based on SF-36 questionnaire by age group of 15-25 years and above and 25 years old in β-thalassemic patients referring to Mofid Hospital showed that there was no significant difference in the dimensions of quality of life of SF-36 scales between the above age groups (Table 6).

Table 6: Comparison of mean percentage of different dimensions of quality of life based on SF-36 questionnaire by age group in patients with thalassemia major referring to Mofid Hospital in 2010-2011

Patients' age group Dimensions of the SF-36 questionnaire	*15-24 years' old standard mean ±) deviation)*	*15 years and older standard mean ±) deviation)*	*Significance level*
Physical function	77/26±4/2	78/25±3/3	0/771
Physical problems	68/33±1/0	68/37±7/1	0/844
Physical pains	73/18±1/1	63/19±3/7	0/102
general health	66/19±7/6	61/17±8/5	0/357
Joy and joy	66/19±9/8	64/22±2/9	0/673
Social performance	77/19±1/5	80/12±2/4	0/841
Mental problems	69/34±5/9	70/20±5/5	0/680
mental health	70/20±8/3	71/18±3/4	0/937

Comparison of GAS scores by age group (15-24 years old and 25 years old) among β-thalassemic patients in Mofid Hospital showed that there was no significant difference in GAS scores between these age groups (P =0 .685) (Figure 7).

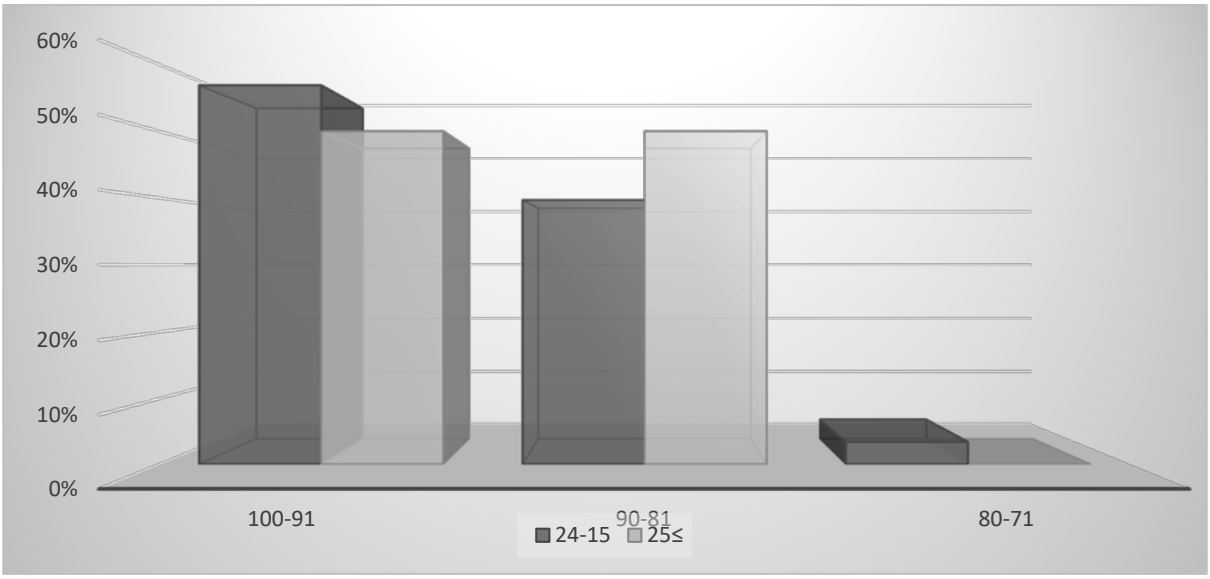

Diagram (7): Comparison of the frequency of general scales of function by age group in patients with thalassemia major referring to Mofid Hospital 2010-2011

The results of comparing the scores of quality of life dimensions of patients with β-thalassemia major were summarized using SF-36 questionnaire based on the age of the first blood transfusion in Table 7. There was no significant difference between the mean scores of different dimensions of quality of life in patients with beta-thalassemia major in the two age groups of the first blood transfusion less than 4 years and 4 years and above (Table 7).

Table 7: Comparison of mean scores of scales of different dimensions of quality of life based on SF-36 questionnaire by age groups beginning of the first blood transfusion in patients with thalassemia major referring to Mofid Hospital from 2010 to 2011

Age of first blood transfusion Dimensions of the questionnaire SF-36	*<4years standard mean ±) deviation)*	*≥ 4 years standard mean ±) deviation)*	*The significance level*
Physical function	75/27±4/9	82/21±0/1	0/479
Physical problems	67/32±5/5	69/36±6/1	0/723
Physical pains	73/18 ±0/4	68/19±3/0	0/272
general health	69/18±0/0	59/20±5/3	0/060
Joy and joy	67/20±3/9	64/18±6/9	0/593
Social performance	77/20±1/4	78/13±8/8	0/923

Mental problems	73/33±0/8	63/38±8/8	0/399
mental health	71/20±9/7	68/18±9/2	0/550

Comparison of GAS in patients with β-thalassemia major in Mofid Hospital based on the age of onset of the first blood transfusion showed that there was no significant difference between the general performance scales and the age at which the onset of the first blood transfusion under the age of 4 years and 4 years or older was not statistically significant (P = 0/108) (Figure 8).

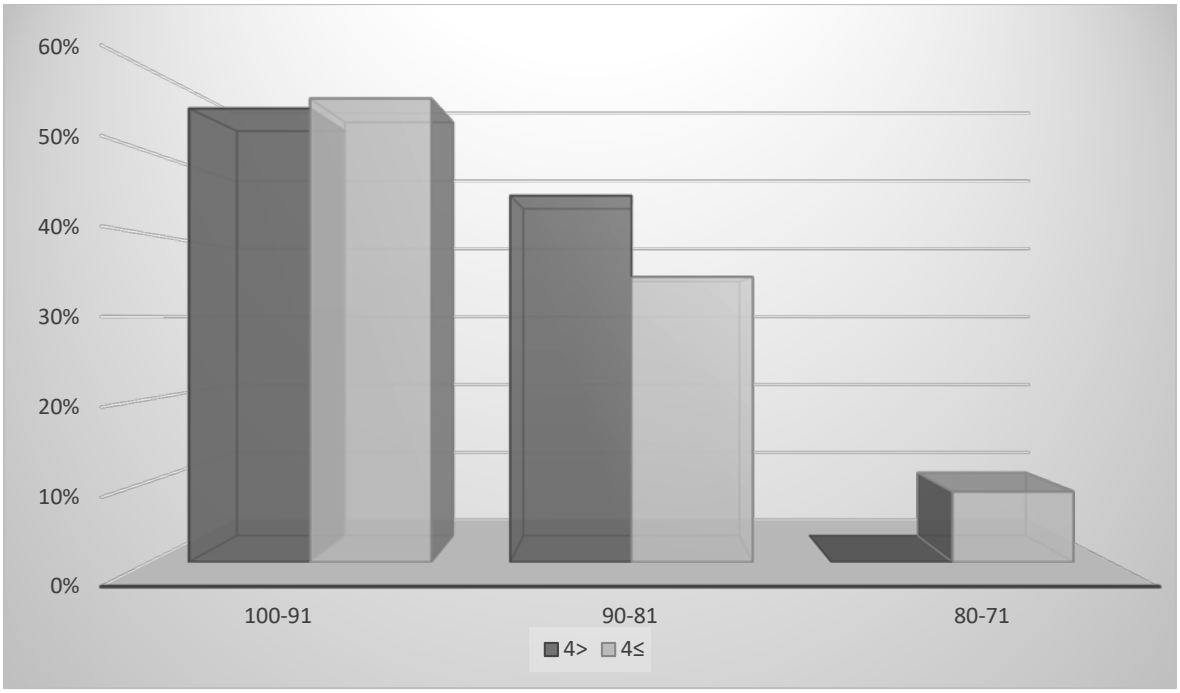

Diagram (8): Comparison of the frequency of general scales of function by the age groups of the first blood transfusion in patients with thalassemia major referring to Mofid Hospital in the years 2010-2011

The mean scores of different dimensions of quality of life in patients with β-thalassemia major referring to Mofid Hospital, based on the SF-36 questionnaire, are summarized in Table 8. The findings showed that there was no significant difference between the different dimensions of quality of life and the presence of related diseases (Table 8).

Table 8: Comparison of Mean percentage of Scores of Scales of Different Dimensions of Quality of Life Based on SF-36 Questionnaire Depending on the presence of associated illnesses in patients with thalassemia major referring to Mofid Hospital in 2010-2011

Associated illnesses Dimensions of the questionnaire SF-36	*Positive standard mean) deviation)±*	*Negative standard mean ±) deviation)*	*Significance level*
Physical function	78/23±5/4	77/±27/0	0/932

Physical problems	70/32±0/0	67/34±5/3	0/816
Physical pains	65/19±5/3	73/17±8/9	0/092
general health	67/23±5/3	65/17±2/5	0/446
Joy and joy	69/20±5/1	65/20±2/3	0/425
Social performance	82/14±5/3	75/19±7/6	0/203
Mental problems	80/33±0/2	66/35±0/4	0/108
mental health	76/19±4/2	68/19±7/8	0/144

Comparison of GAS in patients with β-thalassemia major in Mofid Hospital based on the presence of associated illnesses showed that there was no significant difference between the scales of overall performance and the presence of associated diseases (P = 0.066) (Figure 9).

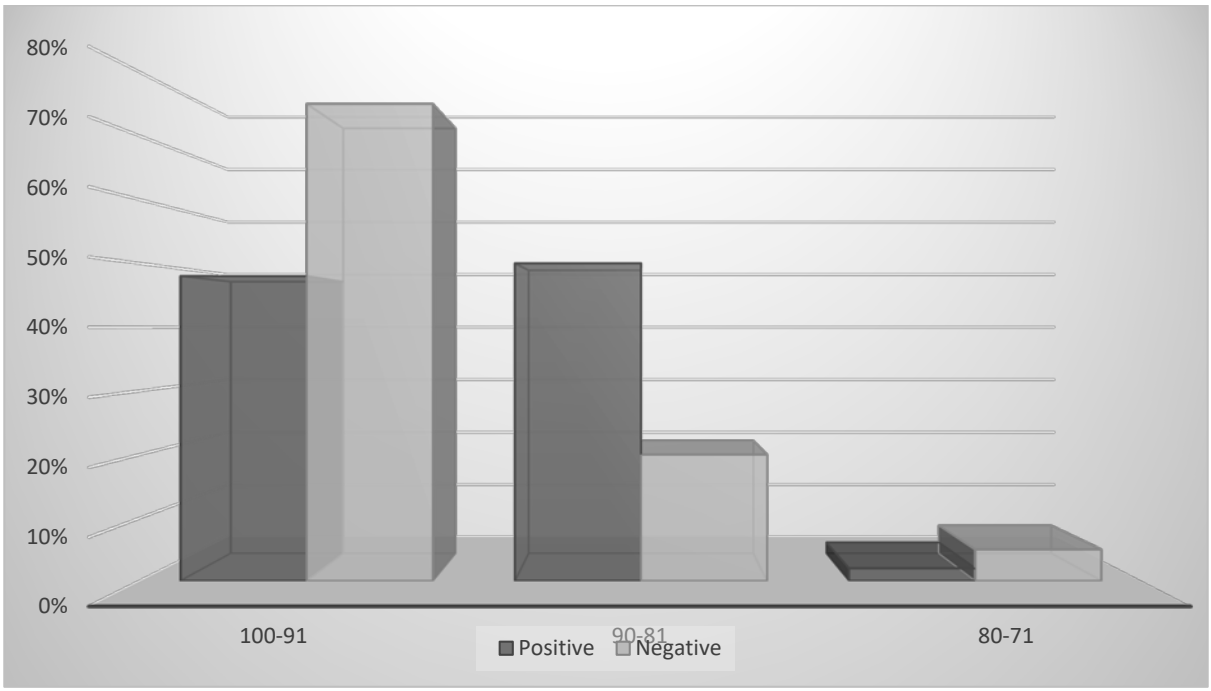

Diagram (9): Comparison of the frequency of total scales of function in terms of the presence of associated diseases in patients with thalassemia major referring to Mofid Hospital in the years 2010-2011

Discussion and conclusion

In the study of Dr. Hadi et al. in Shiraz in 2009, 250 major thalassemia patients found that women had more social problems in terms of social function than men, and stated that this could be justified by the fact that women were inherently more concerned with their problems. And have stronger connections with others and, therefore, have a better social performance than men. In other aspects of quality of life, there was no significant difference between men and women. The mean scores in physical function dimensions and role limitation due to physical problems in the patients group were directly related to age, and the scores of the dimensions were

increased and the findings of this study indicate acceptance of the disease by patient's side or increase in person's self-concept was due to the use of existing therapeutic methods and as a result, reduced facial expression. In the present study, patients with β-thalassemia major had a lower score in terms of overall physical health of their quality of life, especially in terms of physical function and physical pain. They suffered from more physical problems and were able to carry out heavy activities such as running and taking part in sports. They had no power and are limited in performing moderate activities such as light sports, and even had more fatigue than other peers even in lighter activities such as climbing up the stairs and walking more than a few alleys. It is concluded that the low quality of life of these patients in the field of functional performance confirms the complications of this chronic disease, so that hypoxia and hemosiderosis result in endocrine disorders and neurological complications such as mental disorder, spinal cord injuries and involvement 2nerve roots and peripheral nerves and muscular apparatns device Muscle and joint involvement cause particular pain in the ankles, wrists and elbows. The obtained results by M. Tahavorncharoensap [14] and Dr. Hadi's [15], are in agreement with the results of the study of D. shaligram [16] in Bangalore, which was performed on 39 patients with β-thalassemia major. In the present study, β-thalassemia major patients had a better quality of life with regard to the components of mental health and its dimensions including cheerfulness and vitality, social function and role limitation due to emotional problems and mental health than physical dimensions, and social function including family and friends relationship was at the optimal level, and the mental problems in these patients did not affect the following: the time spent working or activity, getting what they want, accurately doing things. In this study, it was concluded that the quality of life of patients with β-thalassemia major in mental health was desirable. Considering that of 70 patients, 58 (82.9%) were in the age group of 15-24 years old and 12 17.1%) were 25 years or older. It seems that the following factors have been effective in improving the mental health of patients: the majority of patients were adolescents and young people and were cared by their parents and still had no problems with life. Strong family and good correlation between the patients themselves have made them better able to adapt to the illness and suffer from less mental problems [17]. Regarding the fact that β-thalassemia major is one of the specific diseases and in Iran our health care services are provided free of charge to this group of patients, reducing the financial burden and not imposing the cost of treatment of these patients on their parents is effective in improving the care of these patients. Since there is a good planning and good follow up in the hospital in terms of the complications of this disease for major thalassemia, the face changes in this disease has significantly decreased, which helps improve the image and acceptance of the disease by the patient and promote the level of patients' quality of life, especially in mental health. Considering that in the overall performance evaluation scale, none of the patients had a score of under 70, had no suicidal thoughts and no anti-social behaviors, it does not seem that these patients have severe psychiatric disorders, although the exact diagnosis of these disorders requires a thorough psychiatric examination. Only 3% of patients had mild functional degradation, which was referred to the psychiatric clinic for further investigation and follow up. Similar to the above results, Dr. Hadi and his colleagues at Shiraz in 2009, did not find any difference between the rate of frustration in the patients with major β thalassemia and their peer control group [15]. Unlike the results of the present study, D. Shaligram and his colleagues in Bangalore in 2007 showed that β-thalassemia major patients had unsatisfactory quality of life in 74% of cases and psychological disorders in 44% and anxiety in 67% and depression in 62% and behavioral problems were reported in 49% of cases (16-15). In the present study, there was no relationship between the presence of associated diseases and the score of SF-36 as well as GAS, which may be due to rapid identification and proper treatment of diseases, so that diseases such as diabetes, hypothyroidism and hypoparathyroidism, and early diagnosis of heart failure and with proper treatment and adequate follow-up of the complications of these diseases are prevented. Contrary to the results of this study, Montarat et al. reported in Thailand in 2010 that the presence of disease complications and the severity of the disease have reduced the quality of life of patients with major β-thalassemia [14].

Conclusion

In this study, patients with thalassemia major were in a good position in terms of mental health. However, in physical health, physical dimensions and physical pain diminished the quality of life in them and may affect the mental health in the second place, causing psychiatric disorders such as anxiety and depression. It seems that the provision of new therapies with the following aims will improve the quality of life of these patients: measures to prevent the development of skeletal disorders and changes in the face of these patients, prevention and timely treatment of transfusion complications, additional treatment with more reliance on oral treatments to prevent pain caused by the injection of iron chelating compounds, appropriate treatment of complications such as diabetes and hypothyroidism and hypoparathyroidism and hypogonadism and short stature and puberty issues, bone marrow graft and transplantation of hematopoietic stem cells, diagnosis and timely treatment of psychiatric disorders in patients with thalassemia major and their family members, and attracting support from various organizations to provide future life for these patients wich is important in terms of education, occupation and marriage in order to improve their quality of life.

References

1. Orkin SH, Nathan DG, Ginsburg D, Thomas L, Fisher DE, Lux SE, et al, editors. Nathan and Oski's Hematology of infancy and childhood. 7th ed. Philadelphia: Saunders; 2009.

2. Sadock BJ, Sadock VA, Ruiz P. editors. Kaplan & Sadock. Comprehensive Textbook of Psychiatry. 9th ed. Philadelphia: Lippincott Williams & Wilkins; 2009.

3. Lichtman MA. Beutler E, Kipps TJ, Seligsohn U, Kaushansky K, Prehal JT, et al, editors. Williams Hematology,7th ed. U.S.A: Mc Graw-Hill Companies; 2006.

4. Michael R, vichinsky E. Thalassemia Syndromes. In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF, editors. Nelson Textbook of Pediatrics. 18th ed. Philadelphia: Saunders; 2007: 2033-7.

5. Rund D, Rachmilewitz E. ß-Thalassemia. N Engl J Med 2005; 353:1135-46.

6. Mikelli A, Tsiantis J. Brief report: Depressive symptoms and quality of life in adolescents with ß-Thalassemia. J Adolesc 2004; 27: 213-16.

7. Telfer P, Constantinidou G, Andreou P, Christou S, Modell B, Angastiniotis M, et al. Quality of life in thalassemia. Ann NY Acad Sci 2005; 1054: 273-82.

8. Old J, olivieri NF. Diagnosis and management of thalassemia. In: Weatherall DJ, Clegg JB, editors. The thalassemia syndromes.4th Ed. New York: Saunders; 2001: 686-722.

9. Politis C.The Psychosocial impact of chronic illness. Ann N Y Acad Sci 1998; 850: 349-54.

10. Moorjani JD, Issac C. Neurotic Manifestations in Adolescents with Thalassemia Major. Indian J Pediatr, 2006 Jul; 73(7): 603-7.

11. 18Montazeri A, Goshtasebi A, Vahdaninia M, Gandek B. The Short Form Health Survey: Translation and Validation study of the Iranian version. Qual Life Res. 2005; 14: 875-82.
12. 19Jafari H, Lahsaeizadeh H, Jafari P, Karimi M. Quality of life in thalassemia major: Reliability and validity of the Persian version of the SF-36 questionnaire. JPGM 2008; 54(4): 273-275.

13. 22Allahyari A, Alhany F, Kazemnejad A, Izadyar M. The effect of family-centered empowerment model on the quality of life of school-age ß-thalassemic children. Iran J Ped 2006; 16 (4): 455-461. (Full Text in Persian)
14. 17Thavorncharoensap M, Torcharus K, Nuchprayoon I, Riewpaibo A, Indaratna K, Ubol B, et al. Factors affecting health-related quality of life in Thai children with thalassemia. BMC Blood Disorders. 2010; 10(1): 1-10.
15. 21Shaffer D, Gould MS, Brasic J,Ambrosini P, Fisher P, Bird H, et al. A children's global assessment scale. Arch Gen Psychiatry 1983; 40 (11): 1228-31.
16. 16Shaligram D, Girimaji SC, Chaturvedi SK. Psychological Problems and Quality of Life in Children with Thalassemia. Indian J Pediatr. 2007 August; 74(8): 727-30.
17. 23Tseng H, Lu JR, Gandek B. Cultural issues using the SF-36 Health Survey in Asia: results from Taiwan. Health & Quality of Life Outcomes 2003; 1: 72-8.