IJBC 2016; 8(4): 108-111

Iranian Journal of Blood & Cancer

Journal Home Page: www.ijbc.ir

ORIGINAL ARTICLE

Health-Related Quality of Life in β Thalassemia Major Children in North of Iran

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ARTICLE INFO

Article History:
Received: 15.08.2016
Accepted: 3.10.2016

Keywords:
β thalassemia major
Health-related quality of life
Physical functioning
Social functioning
Emotional functioning

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ABSTRACT

Background: Advances in treatment of thalassemia major have improved the life expectancy of the patients and therefore their quality of life as other chronic diseases. This study was conducted to assess health- related quality of life in these patients in Guilan province.
Methods: In a cross-sectional study, thirty-one children, aged 8-12 years, with β-thalassemia major were interviewed in Guilan, northern Iran, from January to March 2016. Source of data were health centers of the province and its satellite centers, blood transfusion organizations, general hospitals and private clinics. Health-related quality of life was assessed by using PedsQL™ questionnaire. The Questionnaire was completed at baseline by all patients and their parents. T and Chi-square tests were used as appropriated.
Results: Of the 31 children, 58.1% were girls. Total summary score in children was 75.9±20.1. Physical, Emotional, social, school and psychosocial functioning scores were 70.6±24, 73.3±22.9, 85.9±21, 74.1±21.5, 77.7±19.7, respectively. None of the children underwent splenectomy. Sex, Serum ferritin and hemoglobin levels did not show any association with quality of life in this study.
Conclusion: Although quality of life in these patients was acceptable, HRQOL showed lower scores in comparison to the healthy population. It seems more social and familial support for increasing the quality of life of these children is surely needed.

Please cite this article as: Jafari-Shakib A, Davoudi-Kiakalaye A, Pour-Fathollah AA, Jafari-Shakib R, Mohtasham-Amiri Z. Health-Related Quality of Life in β Thalassemia Major Children in North of Iran. IJBC 2016; 8(4): 108-111.

Introduction

Thalassemia is the most common hereditary anemia in the world with approximately 300 million carriers.[1] This hemoglobinopathy is more prevalent in East Mediterranean region, Southeast Asia and India .[2-5] Thalassemia also is a major health problem in the northern and southern provinces of Iran. Guilan is one of the most prevalent areas of this disease with about 10% of whole thalassemia population .[6]

Although thalassemia major is fatal if not treated appropriately, due to advances in diagnostic methods and treatment measures during the past decade, prognosis and survival rate of these patients has improved significantly.[7-10]

One of the concerns by improving survival of these people as other chronic diseases is their health-related quality of life. Thalassemia is a chronic disease that presents a diverse range of serious clinical and psychological challenges. These challenges not only do affect on the patients' physical, emotional and social functioning but

also their quality of life and relationship with their family.[6] For chronic diseases such as thalassemia, where a cure is not attainable and lifelong treatment is needed, HRQOL is likely to be an essential outcome and the allocation of health care resources seem mandatory.

Numerous studies in different geographical areas on quality of life of this group of patients have shown variable results depending on their living environment, access to medical services, available social and familial supports.[6,11-18]

Thalassemia screening program begun in 1997 in Iran as other countries in the middle-east region resulting in reduction of birth rate of β thalassemia major patients. On the other hand, improving access to health care systems and new treatment modalities have increased life expectancy of these patients. Unfortunately, there is paucity of studies on the quality of life of children suffering from thalassemia major in Iran.[19-20] The objective of this study was to determine the actual situation of health-related quality of life in children with β-thalassemia major in Guilan province.

Materials and Methods

In a cross-sectional study, the following organizations were assessed for access to the data of total number of thalassemic patients in Guilan province. All records of non-communicable disease centers, health centers, hospitals and clinics of hematology were extracted and were merged into a single file, using Excel 2012. A total of 45 patients 8-12 years were found of whom thirty-one were interviewed by trained medical team after obtaining written consent from their parents. Socio-demographic data and history of their disease was gathered from their parents. Last level of ferritin and hemoglobin was taken from their records. PedsQL™ for age 8-12 years was used for assessing quality of life of these children. Persian version of this questionnaire had already been validated.[21] The internal consistency reliability of the PedsQL 4.0 Generic Core Scale approached 0.90 for self-report.[22]

The aim of the study was explained to the children and their parents. Participation in study was optional; therefore, verbal consent was taken from all of them. This questionnaire consisted of 23 questions in Likert scale. The PedsQL 4.0 encompasses the essential core domains for pediatric HRQOL measurement: 1) Physical functioning (8 items), 2) Emotional functioning (5 items), 3) Social functioning (5 items) and 4) School functioning (5 items). In the "PedsQL™ Generic Core Scales", for ease of interpretability, items are reversed scored and linearly transformed to a 0-100 scale; the higher points reflect the higher quality of life. To create the psychosocial health summary score, the mean is computed as the sum of the items divided by the number of items answered in the emotional, social, and school functioning scales. The "Physical Health Summary Score" is the same as the "Physical Functioning Scale Score". To create the Total Scale Score, the mean is computed as the sum of all the items divided by the items answered over all the scales.[22]

Data analysis was done by SPSS software, version 18 using description statistics, T and Chi-square tests.

Results

Forty-five children aged 8-12 years with β-thalassemia major living in Guilan province, northern Iran, were enrolled. Of these, 31 patients participated in this study during study period. 18 children (58.1%) were girls and most (64.5%) lived in rural areas. The most common educational level of the parents was diploma or lower. All the patients were receiving iron chelating medications. None of the patients had undergone splenectomy or bone marrow transplant. Age at diagnosis was from 2-6 months old. Mean household income was 270±109 US dollars (range: 67-467 US dollars). The average number of blood transfusions per year was 12 times. Mean hemoglobin and ferritin levels were 8±0.9 g/dl (5.9-9.7 g/ dl with median level of 8.1 g/dl) and 1377.5±990 ng/ml (250-4019 ng/ml with median level of 1100 ng/ml), respectively.

Total score in children was 75.9±20.1. Physical functioning, Emotional functioning, Social functioning, and School functioning and "Psychosocial Health Summary Scores" were 70.6±24, 73.3±22.9, 85.9±21, 74.1±21.5, 77.7±19.7, respectively (table 1). Sex, serum ferritin and hemoglobin levels did not show any association with quality of life of the patients.

Discussion

Thalassemia is the most common hereditary anemia in Iran. Treatment measures include allogeneic hematopoietic transplantation if a suitable donor is available or lifelong regular transfusion, iron chelation, splenectomy in occasional cases and supportive measures in terms of growth and nutrition of the patients.

With major improvements in treatment of thalassemic patients in recent years, survivals of the patients have increased and their quality of life has become a major concern for health policy makers and physicians. Studies in different geographical areas have shown that quality of life of the patients highly depend on socio-economic status, living environment, access to medical services and social and family supports.

Table 1: Mean PedsQL 4.0 Generic Quality of Life Scores in patients

Domain	Mean±SD	Median	Min-Max
Physical Functioning	70.6±24	71.8	0-100
Emotional Functioning	73.3±22.9	80	0-100
Social Functioning	85.9±21	90	0-100
School Functioning	74.1±21.5	75	0-100
Psychosocial Health Summary	77.7±19.7	78.3	0-100
Total summary score	75.9±20.1	77	0-100

Previous study illustrated that children above 6 years of age can tell their own health status.[12] HRQOL in our patients indicated higher scores and better conditions in comparison to studies of Ismail in Malaysia,[12] Cheuk in Hong Kong,[23] and Garaibeh in Jordan.[16] However, the scores achieved in our study were similar to other studies.[5,11,17,24] Based on our results physical and school functioning had lower scores than emotional and social functioning. Frequent absenteeism and low physical performance may be the causes of negative impact of this disease on children's quality of life. It seems that more educational programs for teachers and students could make them more familiar with this disease and asking teacher to spend more time for these students have an important role in improving their situation. In our study, sex did not have any impact on quality of life, but other studies have shown other results in which women have a lower scale of quality of life.[25,26]

There are also limited studies which show the influence of household income as a contribution factor in quality of life.[17,27]; however, our study did not confirm it. Inconsistent with finding of other studies;[5,28,29] serum hemoglobin levels did not show any association with quality of life of the patients. Appropriate control of the disease in our patients may be the reason for almost acceptable quality of life as more than half of our patients had hemoglobin level of ≥8 g/dl. Of course small sample size could be another reason for this inconsistent.

Consistent with a previous study in Thailand[5] serum ferritin level had not any association with quality of life of the patients although many other studies had shown conflicting results.[8,30,31] Lower age of patients could explain this controversy that short term iron overload did not cause significant complaints and complications. Meanwhile in this study about 44% of patients had serum ferritin level less than 1000 ng/ml.

It seems that over the economic situation, cultural status and acceptance of disease with other member of family are important factors in quality of life.

Conclusion

This was the first study to provide evidence of HRQOL of children with β-thalassemia major in Iran. We found lower scores compared with healthy population. Our study revealed this population need more attention from Health Ministry, Ministry of Education, school authorities and the society to provide them better quality of life.

Acknowledgement

The authors express their gratitude to Iranian Blood Transfusion Organization for their financial support.

Conflict of Interest: None declared.

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