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Study of quality of life from the perspective of parents and children with thalassemia major

Mostafa Ali Mehr[1], Hoda Kalhorpour[2], Vahid Shirani[3], Maryam Seyed-Nezhad[4]✉, Ali Kord[5], Mehdi Zainivand[6]

Objective: **Thalassemia major is a genetic disease with symptoms of a severe and chronic anemia. This disease is one of the most prevalent chronic genetic diseases in the world and the Iran. About two hundred million people are afflicted with this disease all over the world. The tools for evaluation of life quality enable us to have a closer look at the impact of disease on children's life quality. This study is aimed at studying the quality of life from the perspective of the parents and children with thalassemia.** ***Method:*** **Thalassemia major is a genetic disease with symptoms of a severe and chronic anemia. This disease is one of the most prevalent chronic genetic diseases in the world and the Iran. About two hundred million people are afflicted with this disease all over the world. The tools for evaluation of life quality enable us to have a closer look at the impact of disease on children's life quality. This study is aimed at studying the quality of life from the perspective of the parents and children with thalassemia.** ***Findings:*** **The average score of quality of life for these children was 41.01±22.38 from their perspective and 40.63±21.04 from their parent's perspective. From the perspective of the children and their parents, there was a significant difference between the average scores of quality of life both in general and in other components of physical, social, emotional, and school performance (0/0001<P). The average score reported by the parents in physical and social components was higher than that reported by the children, and in other components, this score was lower than the score reported by the children. There was no significant statistical difference between quality of life for the girls and boys. In the component of school performance, rural children had a higher quality of life than urban children (0/05<P).** ***Conclusion:*** **Due to the parents' important role in child care, it is suggested for the thalassemia-related authorities to consider the parents' perspectives about improvement of children's quality of life in care planning and provide the parents with the opportunity of participating in planning for child care to promote the children's quality of life.**

INTRODUCTION

Thalassemia major is one of the most prevalent chronic genetic diseases in the world and Iran. About two hundred million people are afflicted with this disease all over the world. Thousands of children are born with thalassemia annually, and the burden of this disease is high particularly in developing countries [1]. Thalassemia major is a chronic genetic hematologic disease caused by lack of synthesis in one or more polypeptide glob in chains which is transmitted from a generation to another based on Mendelian genetic rules [2] and is non-communicable diseases [3].

This disease is observed with the symptoms of severe and chronic anemia, lack of proper growth, big spleen and liver, bone disorders especially visible changes in head and face bones with change of look [4]. Geographically, thalassemia is Mediterranean disease and is more prevalent in west Asian and central Asian countries such as Turkey, Iran, Burma, Thailand, Vietnam, and Columbia. World Health Organization has recognized thalassemia as the most prevalent chronic genetic disorder among 60 countries of the world and affects the lives of 100000 children annually [5].

In Iran, thalassemia is one of the most prevalent genetic diseases. According to statistics of the foundation for special diseases, 22000 people are afflicted with thalassemia major in the country [6]. In Iran, thalassemia is more prevalent in the margin of Caspian Sea, Persian Gulf, and Oman Sea including Mazandaran, Gilan, Khuzestan, Fars, Bushehr, Hormozgan, Sistan & Baluchestan, and Kerman. In Iran, prevalence of the gene based on geographic area is different and between 3-4 percent [7].

Before applying the treatment method of blood injection, these patients did not have a long life and most of them died in the first decade of life. However, they got a longer life with blood injection in every 3 to 4 weeks and keeping hemoglobin stable and their physical appearance especially their head and face was significantly improved, and also, their growth disorder was decreased [8]. Over the past decade, many advances in treating the patients with thalassemia major have significantly improved their survival. However, there some problems in managing this chronic disease such as iron deficiency with iron chelate treatment, endocrine problems, hepatitis C, and the related social-mental

[1]PhD. Student in Health Services Management, Dezful University of Medical Sciences, Dezful, Iran; [2]Master Student in Demography, *Dezful University of Medical Sciences, Dezful, Iran*; [3]General Dentist, Dezful University of Medical Sciences, Dezful, Iran; [4]PhD. Student in Health Services Management, Department of Health Management and Economics, School of Public Health, Tehran University of Medical Sciences, Tehran, Iran; [5]PhD. Student in Health Services Management, Alborz University of Medical Sciences, Karaj, Iran; [6]Master Student in governmental management, Ilam University of Medical Sciences, Ilam, Iran;

✉*Corresponding author:* [4]PhD. Student in Health Services Management, Department of Health Management and Economics, School of Public Health, Tehran University of Medical Sciences, Tehran, Iran

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effects. The children with this disease suffer from this condition and their social relations are significantly decreased with a feeling of isolation [9].

The results of the studies show that parents having children with chronic diseases experience some degree of frustration and stress, especially when they find out that their children will be afflicted by this disease until the end of their lives. Most of these parents have vulnerable personalities, because they think that they have had a role in their child's illness, or because they cannot care for their child well. In some cases, the family's identity is lost and the parents got depressed [10].

Measuring life quality-related health is a multidimensional concept which is focused on the effect of disease and its treatment on the individual's health. Measuring life quality-related health includes recording patients' perspectives about the disease and their treatment, their feeling of need to receive healthcare, and their priorities for the results of the treatment and disease [11]. There is a mutual relationship between illness and quality of life especially in patients with chronic disease who are faced with various problems during the life stages, and physical disorders and physical symptoms have a direct effect on all aspects of quality of life [27]. The main purpose of treating these patients is to strengthen quality of their lives through reducing the effects of the disease, and they should not have a low quality of life necessarily [12]. Some of the researches show that 80 percent of the people afflicted with thalassemia major have at least one psychiatric disorder. The most prevalent psychiatric disorders reported in these patients or their family members include having a bad mental image of their body, anxiety, depression, physical disorders, and inability to control their anger properly. Also, these patients have various problems in their social and educational activities [13]. The previous studies on the adults with thalassemia showed that different treatment and culture do not have any effect on quality of life. Other studies have shown that age and the related effects are predictive of health related quality of life in children with thalassemia, while age, ethnicity, and family do not have any effect on the life of these patients' lives [14]. A study in India showed that thalassemia is very stressful and the patients face with many physical, mental, and social problems. In their study, Kanatan et al (2003) reported 60% education problems, 20% social interaction problems, 24% feeling of being different, and 31% anxiety in children with thalassemia [15]. In the study by Ghanizadeh et al, the most prevalent mental disorder among these patients is depression and anxiety. They reported 1.8% attention deficiency, 9% bipolar disorder, 49% depression, 62.7% irritability and anger, 43% thinking about death, and 27.3% thinking about suicide [16]. In the study conducted by Esmaeil et al (2006), it was found that the scores of physical, social, and school performance are significantly lower in the patients with thalassemia than healthy children.

The children and adolescents with thalassemia have the most problems of life quality in physical and emotional aspects. Special support by the health authorities, school, and community is necessary to increase the quality of their lives. Therefore, planning for improving mental health, participating in social activities, and consulting on educational psychology besides clinical and treatment services can lead to increased quality of their life [12].

The parents of children with chronic disease also suffer from long term care and the community's reactions to their child's disease and these can lead to stress and mental-social problems for the parents and as a result, decreased general health among them. Parents decreased general health influences the children's life quality and makes the parents consider their child's health very vulnerable. It makes the parents too concerned about protecting their child and making the child restricted and consequently, it leads to negative effects on the child (11 new). Since there has been few researches on comparing the perspective of the children with thalassemia major with their parents' perspectives in Iran, we decided to conduct the current research aimed at studying the perspective of the children with thalassemia major and their parents about quality of life. The findings can be sued for planning a proper and comprehensive program to improve the quality of life for these children and reduce the effects of the disease on their physical and mental health and take a step toward preventing the diseases and guaranteeing the health of the children with thalassemia major.

METHOD

In this descriptive-analytical study, 150 children with thalassemia major with the age of 6-15 years old who had referred to Samen-al-Hojaj special diseases treatment center in Kerman with one of their parents were selected by convenient sampling method. The criterion for leaving the study was unwillingness to cooperate for filling the questionnaire.

Children's life quality questionnaire (PedSQL) that considers the children and their parents' perspectives was used (child and parent report). Each of these two questionnaires contained 23 questions in physical (8 items), emotional (5 items), social (5 items), and school performance (5 items) components. Scoring was done by Likert five grade scale ranging 0-100. The score of 100 indicated the least problems and the highest quality of life. In the study conducted by Warney et al (2001), Cronbach's alpha that is an indicator of reliability index, for this questionnaire was equal to 0/88 from the children's perspective and 0/90 from the parents' perspective [17]. Also after the translation, scientific validity was approved by pediatricians of Kerman through content validity, and the Cronbach's alpha was obtained as 0/84 and 0/88 respectively form the viewpoints of the patients and parents. The questionnaires were distributed among the children and the parents by the researcher. The data was analyzed by SPSS 18 statistical software, descriptive statistics, independent t-test, variance analysis, and Mann-Whitney test.

FINDINGS

The studied children were in the range of 5-16 years old with an average of 11/61±2/77 years. 54/7 percent of the children were girls and the remaining were boys. 63/3 percent of the families lived in the cities. 30.7% of the families had four or more children and 15.3% of the families had only one child. 77.3% of the families had a child with thalassemia major and 22.7% of the families had 2 children with thalassemia major. 38% of the fathers and 36.7% of the mothers had lower education than diploma. In this research, respectively 36% and 35.3% of the fathers were self-employed and worker. 89.3% of the mothers were housewives.

From the perspective of the children and the parents, there was a significant statistical difference between the average scores of life quality both in general and in physical, emotional, social, and school performance components (<P 0.0001). The average score reported by the parents in physical and emotional performance was higher than that reported by the children and in other components this score was lower than that reported by the children (Table 1 and chart 1).

T test showed that there is no significant statistical difference between quality of life for the girls and boys (Table 2 and chart 2). Also, according to Mann-Whitney test, quality of life for rural children was higher than urban children in school performance component (<P 0/05); (Table 2 and chart 3).

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Table 1 parent in physical and emotional performance

Dimensions of quality of life	Children	Parents
	Standard deviation±Average	Standard deviation±Average
Physical function	*37.41±26.02	37.7±25.11
Emotional function	*45.76±27.56	45.33±26.32
Social function	*46.86±23.67	47.33±24.17
Performance at school	*36.16±27.83	33.93±25.59
Total	*41.01±22.38	40.63±21.04

**<P-value 0/0001 compared with the parents*

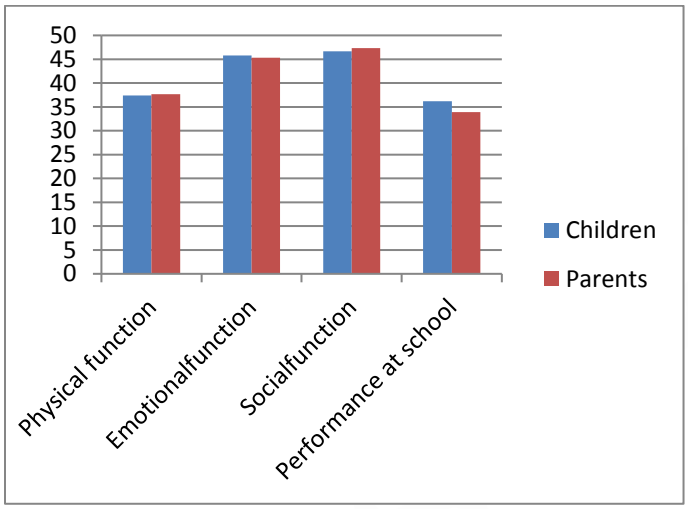

Chart 1 Quality of life for children with thalassemia major from the perspective of the children and their parents

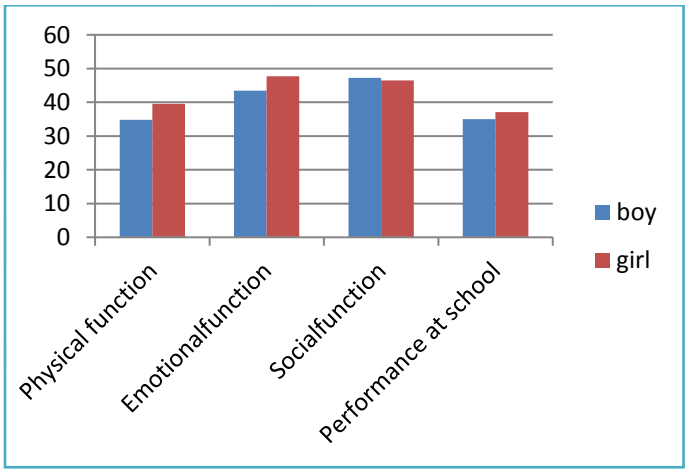

Chart 2 Comparison of the average quality of life for children with thalassemia major based on gender

Table 2 Comparison of the average quality of life for children with thalassemia major based on gender and residence

Dimensions of demographic characteristics / quality of life	Sex		Address	
	boy	Girl	rural	Urban
	Standard deviation±Average	Standard deviation±Average	Standard deviation±Average	Standard deviation±Average
Physical function	34.83±26.67	39.55±25.44	43.35±30.26	33.98±22.69
Emotional function	43.45±27.45	47.68±27.67	50±29.78	43.31±26.04
Social function	47.27±23.50	46.52±23.95	51.36±27	44.26±21.23

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Performance at school	35±28.69	37.13±27.25	45.63±30.94	*30.68±24.4
total	39.45±22.32	42.31±22.49	47.03±26.64	37.52±18.79

<P-value 0/05 compared with the rural community

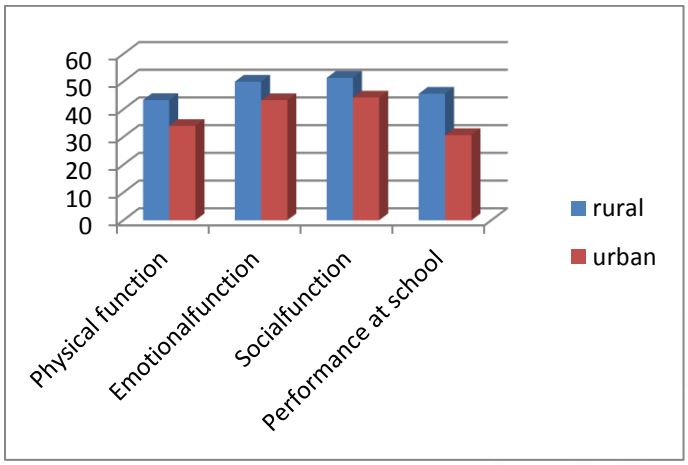

Chart 3 Comparison of the average quality of life for children with thalassemia major based on residence

DISCUSSION

In this study, the four aspects of quality of life (physical, emotional, social, and school performance) from the perspectives of the children with thalassemia major and their parents were compared. From the perspectives of both the children and their parents, school performance component was accompanied by the most problems. Also, the parents reported their children's life quality both generally and in emotional and school performance components lower compared with the perspective of the children. Since most of the parents having children with chronic and especially genetic diseases blame themselves for their child's illness and also due to the pressures of caring for these children, it is obvious that these parents always report the quality of these children's life lower than the children themselves.

In a study aimed at studying the quality of life for the patients with thalassemia and the people caring for them, Zare indicated that the total average of life quality aspects was equal to 70.25±17.17 for the patients and 67.44±16.59 for their parents, and there was a significant difference between the patients and their parents only in social performance component [2]. In a study on children with asthma, Moridi indicated that the parents reported the quality of life lower than the children [18]. In a study on 96 children with thalassemia major and 235 healthy children, Ismail indicated that quality of life for the children with thalassemia was significantly lower than the healthy children [11]. In a study aimed at comparing quality of life for children with cancer from the perspective of the children and the parents, Valizadeh indicated that the children and both parents reported the quality of life higher than medium in psychiatric, family and social aspect. Also, the score of quality of life reported by the parents was lower than the score reported by the children [19]. In a study on 39 children with thalassemia, Shaligram indicated that 44 percent of the children had mental problems and 74 percent of them had an unfavorable quality of life. Anxiety symptoms (67%), emotional problems especially depression (62%), and behavioral problems (49%) were the main findings of this research [20].

Lindsey conducted a study on 364 children and adolescents with elbow disorder and 164 parents. The results of the study showed that the parents reported their children's physical and social performance much lower than the children themselves (P<0.001) [21]. In a study on 178 children and adolescents with thalassemia major and their parents, PishkarMofrad indicated that the parents had reported their children's life quality lower than the children themselves [22]. In a study on 20 adolescents with thalassemia major to determine their mental health by general health questionnaire including four areas of physical symptoms, anxiety, social performance disorder, and depression, Pooraboli indicated that in all the four areas, the percentage of healthy people was far less than people with mental health disorders [15]. Panepinto conducted a study on 53 children with sickle cell anemia. In this study, the parents reported the quality of life lower than the children both in general and in physical performance, behavior, and self-esteem [23].

In a study on 40 children with thalassemia major and 40 classmate children, Khaledi indicated that quality of life in the four components of physical, emotional, social, and school performance, the children with thalassemia were at a significantly lower level than the children in the control group [24]. Bullinger and Von Mackensen showed that quality of life in emotional and social aspects for children with hemophilia was different from the perspectives of the children and the parents, and these two aspects were at a worse condition from the parents' perspective [18].

The results of the current research were similar to the above studies and it suggests that chronic diseases in children such as thalassemia lead to a significant impact on the patient and the relatives' life. Besides physical complications, the other issue that is paid more attention is the concerns and emotional problems that suggest that having a child with a chronic disease is an everyday stress for the family.

There was not any significant statistical difference between quality of life for the girls and boys with thalassemia major. In a study on 250 patients with thalassemia major and a 100-people control group, Hadi showed that quality of life for the patients with thalassemia in terms of physical health and the related aspects was lower than the average scores of life quality in the control group. However in mental health and the related components, there was no significant statistical difference between the two groups. Also, the female patients received higher scores of social performance than male patients [25]. In a study on the children with diabetes, Faulkner indicated that girls with diabetes have a lower quality of life than boys with diabetes [26]. The results of the current

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study are not consistent with the above study and suggest no difference in quality of life for the girls and boys with thalassemia major. This lack of difference can be attributed to the age range of these children because in this age group, children depend on their parents to do their physical activities and they have not get a social and emotional understanding yet. Also, based on Mann-Whitney test, the average score of school performance for rural children was higher than urban children, and this is justifiable regarding low expectations of rural children than the urban children. In a study on 178 children and adolescents with thalassemia major and their parents, Pishkar Mofrad indicated that rural children had a lower quality of life than urban children [22]. In a study on 39 children and adolescents with thalassemia major, Alavi indicated that the average score of all the aspects of quality of life for the rural children was higher than that of the urban children, and as a result, rural children reported a lower quality of life [6].

CONCLUSION

According to the results of this study, quality of life in physical, emotional, social, and school performance components is lower than average from the perspectives of both the children and their parents. Therefore, there is need to the presence of experienced psychiatric counselors in Kerman special diseases treatment centers for education of life skills and dealing with emotional and social problems and supporting the patients and their parents.

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Ethical Considerations
Written informed consent was obtained from the patient, and the study was approved by the Kerman University of Medical Sciences Ethical Committee and conducted in accordance with the ethical guidelines of the Declaration of Helsinki (IR.KMU.ACRS.REC.1396.1055).

Conflict of Interests
There is no conflict of interest among the authors.

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