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Investigating Challenges Facing the Improvement of Health Related Quality of Life in Iranian β-Thalassemia Major Patients: A Qualitative Study

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Abstract

Background: The complexity of thalassemia and its treatment process has a significant impact on the quality of life of thalassemia major patients. The aim of this study was to investigate the challenges of improving health-related quality of life in β-thalassemia major patients.

Methods: A qualitative content analysis was performed at the thalassemia center in Mashhad, Iran. Semi-structured interviews were performed from January 2019 to May 2020, 25 interviews were performed with 23 participants (15 patients aged ≥18 years, 3 family members and 5 caregivers) who were selected by purposeful sampling.

Results: After analyzing the data, the challenges of improving health-related quality of life in β-thalassemia major patients were categorized into five major themes, including I. living activity, II. opportunity and motivation, III. adaptation, IV. received healthcare, V. psychological and social support.

Conclusions: Age increase among thalassemia patients is associated with increased physical, psychological and social complications and treatment costs that lead to a reduced HRQOL. By increasing age, it is necessary to arrange some programs for their presence in the community and to take particular measures for employment, education and marriage of thalassemia major patients. As life span increases, the treatment of these patients reaches the cost-effectiveness threshold, so policy-making is critical for screening the complications of the disease. Along with providing a perfect treatment environment, the accessibility of appropriate laboratory and equipment for measuring iron deposition should be the priorities for health care system of countries, so that thalassemia patients may experience fewer complications and higher HRQOL in their life.

Key Words: Barriers; β-thalassemia major; Caregiver; Health related quality of life; HRQOL; Qualitative research.

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1- INTRODUCTION

Iran is one of the countries located in the thalassemia belt. By 2019, almost 18600 patients with thalassemia major have been detected in Iran (1), which ranks first in the world in terms of the ratio of thalassemia patients to the total population of the country (2). Although the northern and southern provinces of Iran have a large number of thalassemia major patients or the carriers, in immigrant provinces such as Razavi Khorasan, the birth of these patients is significant. Thus, it is necessary to conduct thalassemia related research studies for national decision making (3). Patients with beta thalassemia major need regular blood transfusions due to severe anemia to survive and grow properly (4); over time it leads to iron overload, and to prevent complications, the patients need iron chelator medications (5). Vizza's study (2018) that examined the caregiver experiences of thalassemia adolescents transitioning to adulthood showed that as the patients' age increased, they experienced more needs for hospitalizations, health services and treatment costs. In addition, the occurrence of complications led to work ban for these patients and brought them about numerous psychological, economic and social challenges; and led to a reduced health-related quality of life (HRQOL) in these patients (6). A study by Waheed et al. (2015) showed that HRQOL in thalassemia major patients was highly dependent on national thalassemia care programs. Therefore, it is essential to pay attention to the management and care programs of these patients in the healthcare system of countries with higher prevalence of thalassemia (7). National thalassemia prevention program has been started since 1997 in Iran and it has been able to prevent about 80% of the birth of babies with thalassemia major and 53% of the marriages of two thalassemia gene carriers (3); and their raw mortality rate in 2017 was less than 4.7 per 1000 people. A meta-analysis conducted by Arian et al. (2019) showed that HRQOL score of thalassemia major patients in Iran was lower than the general population and the mean age of these patients in Iran was almost 5 years below other countries (1). As mentioned, Iran has the most successful program to reduce the birth rate of thalassemia babies in the region due to primary and secondary national thalassemia prevention program, but a comprehensive program to improve HRQOL of the patients has not yet been provided (7). Over the last few years, development of therapeutic and pharmacological methods, especially the production of oral iron chelator medications, has increased treatment adherence in these patients and has led to an increase in their lifespan; therefore, in parallel with the increase in life expectancy, planning for HRQOL promotion should be included in the management programs of the disease (1). Despite extensive efforts to provide health care services for thalassemia patients, there are still some deficiencies in the quality of caregiving provided to these patients that directly reduce their HRQOL. Accordingly, it seems necessary to identify barriers of HRQOL promotion, and to make fundamental changes and new strategies to remove the barriers (8). This strategy should be rational and based on the patient's ability and potentials of the health care system. It should also be able to improve the patient's HRQOL and reduce his/her dependency on health system. Furthermore, it should consider cost saving, reducing the burden of the disease on the patient and society (9). In addition to identifying the factors influencing HRQOL in chronic patients, Sedlar et al. (2020) considered that regular evaluation of these factors was critical, because these actions were necessary to take in order to improve the functioning of health care system towards standardizing

the quality of care through a patient-centered approach in chronic patients (10). Ford et al. argued that for making any change in management systems aimed at empowering patients and improving their HRQOL, potential barriers should first be identified (11). Developing targeted protocols to meet patients' needs, experiences, and interests is still a pivotal challenge that needs to be addressed through proper research approaches based on patients' life experiences (10, 11). Thalassemia is a common genetic disease in Iran and it is necessary to identify beneficial approaches to the management of the disease in order to promote HRQOL in these patients (1). Identifying these approaches _ if they are consistent with the existing reality and based on the patients' life experience_ can help managing care and self-care interventions, improving health outcomes, and promoting HRQOL (7). Hereupon, the present qualitative study investigates the challenges of promoting HRQOL in beta thalassemia major patients in an Iranian population.

2- METHODS

2-1- Aims

The present qualitative study investigates the challenges of promoting HRQOL in beta thalassemia major patients aged ≥ 18 years from Iranian population.

2-2- Study Design and Participants

In this qualitative study, semi-structured interviews and purposeful sampling were used. Participants were selected from thalassemia major patients over 18 years of age, who referred to Mashhad Thalassemia Center in Iran for blood transfusion; they were able to communicate properly. The utmost variability in sampling was considered in terms of different levels of adherence to iron chelator drugs as well as various sociological, economic, and educational situations of the participants. Purposeful sampling is widely used in qualitative research for the identification and selection of information-rich cases for the most effective use of limited resources related to the phenomenon of study. So participants were selected from various groups. Besides, family members of patients and health care professionals working in thalassemia care centers were interviewed as rich cases and resources related to the phenomenon of study.

2-3- Data Collection

This study is a part of a large study as a PhD thesis in nursing with the qualitative content analysis design. It was conducted from January 2019 to May 2020 at a thalassemia center in Mashhad, Iran. Some of the analyses related to the challenges of promoting HRQOL among patients are presented in the present study. At first, the objective of the study was explained to the participants by phone or in person. All interviews conducted by the first author (MA), who knew qualitative interview techniques. A semi-structured interview guide (**Table 1**), was prepared by the authors based on the relevant literature (10, 12).

The guiding questions were designed to gather similar data from each participant and to provide a structure for discussion during the interviews. Additional questions were used for clarification and reflection on the emerging issues raised by participants, along with exploring their experiences. Interviews were conducted until the data were saturated. Saturation occurred when previous data were constantly repeated and no new data were obtained. This happened after 25 interviews with 23 participants. Two participants were interviewed twice. All interviews were conducted in Farsi and face to face at the most appropriate place (in thalassemia center in Mashhad, Iran), date and time for the participants. Interviews were recorded. The interviews lasted 30 to 45 minutes. All interviews were recorded by digital voice recorder (Sony MP4-MP5 Made in Japan). The

researcher used written documents, newspapers, pictures, videos, computer programs related to the subject as other sources of data which were taken from the context of the study, as well as informal interviews and observations of documents stored in the archives of Thalassemia Association of Razavi Khorasan, Iran. This data gathering was like a field interview or fieldnote.

Table-1: Dimensions and some of the questions used in the interviews (10, 12)

Questions about the disease	• Tell me about your disease and how your parents found out you had it? • How do you manage your condition, for example dealing with symptoms, specific tasks–taking medication, managing appointments, etc.? • What are the things which make it easier or harder to take care of yourself/ your patients? • What strategies are effective in taking care of yourself/your patients?
Questions about psychological and social support	• How does this disease affect your relationship and communication with relatives/friends/family? • What do you do if you/your patients feel down, frustrated, or anxious?
Questions about the received healthcare in an outpatient thalassemia clinic	• Can you explain your experience as a thalassemia patient in your encounters with the healthcare team (for example, nurses or physicians)? • How do you receive information from healthcare professionals? • What educations and instructions do you receive about taking care of yourself / your patients? • What are the things helping you better fit the educations and instructions into your normal activities? • How well and comfortable can you talk to your healthcare team about your/patient's health concerns?

2-4- Ethical Consideration

The protocol of this study has been approved by the Ethics Committee of the institution in which the first author (MS) worked (decree code: IR.SEMUMS.REC.1397.244). Permission to enter the research area was obtained from the relevant organizations before data gathering. Moreover, the objective of the study was clearly explained to the participants, and before the interview, the informed consent form was signed by those who were willing to participate in the study. The names and confidentiality of the participants' information were guaranteed. Participants were told that they could withdraw from the study at any time without penalty. Prior to the interviews, informed written and verbal consent were obtained from the participants to record the interviews. Participants were assured that the collected data would be analyzed and published anonymously.

2-5- Data Analysis

Data analysis was carried out in accordance with the steps recommended by Elo and Kyngås (13-15). After each interview, the researcher immediately transcribed it into written texts, and each interview was exactly read for several times to gain an overall comprehension of its content. Meaning units were then

identified and the initial codes were extracted. Next, similar initial codes were categorized into more comprehensive categories and sub-categories. Based on the study's objective, the perceived concepts were categorized into three groups (Both of them-Facilitator-Barrier) and the participants' opinions about them were re-evaluated. MAXQDA 20 software was used to analyse the data.

2-6- Rigor

The four criteria of transferability, credibility, dependability, and conformability were used for rigor (16). Credibility was ensured through member- and peer -checking, long-term engagement, and maximum variations in sampling. Concerning the long-term engagement, the main researcher (MA) engaged with the participants in the thalassemia center through spending enough time with the patients, their families and healthcare providers to explore this phenomenon from different perspectives. For member checking, summary information of the findings was reported to some participants to ensure that their experiences were exactly reflected. The data analysis process and findings were also checked with two qualitative researchers who were not the members of the research team; and they approved the coding and analysis process. The study process and findings have been fully described and recorded to facilitate audit trail (17). Transferability is ensured through providing a rich explanation of the actions taken during the data collection and analysis phases (16).

3- RESULTS

23 participants included 15 patients with mean ages of 27.75 ± 7.9 years in women and 27.42 ± 56 years in men, 3 family members of patients with a mean age of 36 ± 2.8 years and 5 caregivers including 2 nurses, 2 physicians and 1 social welfare agent with a mean age of 41.75 ± 5.47 years. The demographic data of the participants are mentioned in **Table 2**.

Table-2: Demographic information of the participants

Participants	Number, Gender	Age, mean (standard deviation), y	Level of education n (%)		Occupation n (%)	
Patient with beta-thalassemia major	7 males 8 females	27.42±5.6 27.75±7.9	High school diploma Associate's degree Bachelor's degree Master's degree	9(60%) 2(13%) 3(20%) 1(7%)	Student Housewife Permanent job Not permanent job	3(20%) 2(13%) 3(20%) 7(47%)
Family member	3 females	36±2.8	Primary school Bachelor's degree Master's degree	1(33.3%) 1(33.3%) 1(33.3%)	Housewife Teacher	1(33.3%) 2(66.6%)
Caregivers	4 females 1 male	36.5±4.58 47	Bachelor's degree Specialist physician	3(60%) 2(40%)	Nurse Psychologist Physician	2(40%) 1(20%) 2(40%)

From the data analysis, five contents were obtained, including: I. living activity, II. opportunity and motivation, III. adaptation, IV. received healthcare, V. psychological and social support (**Table 3**). **Fig. 1** shows barriers and facilities through Ishikawa (fishbone) diagram.

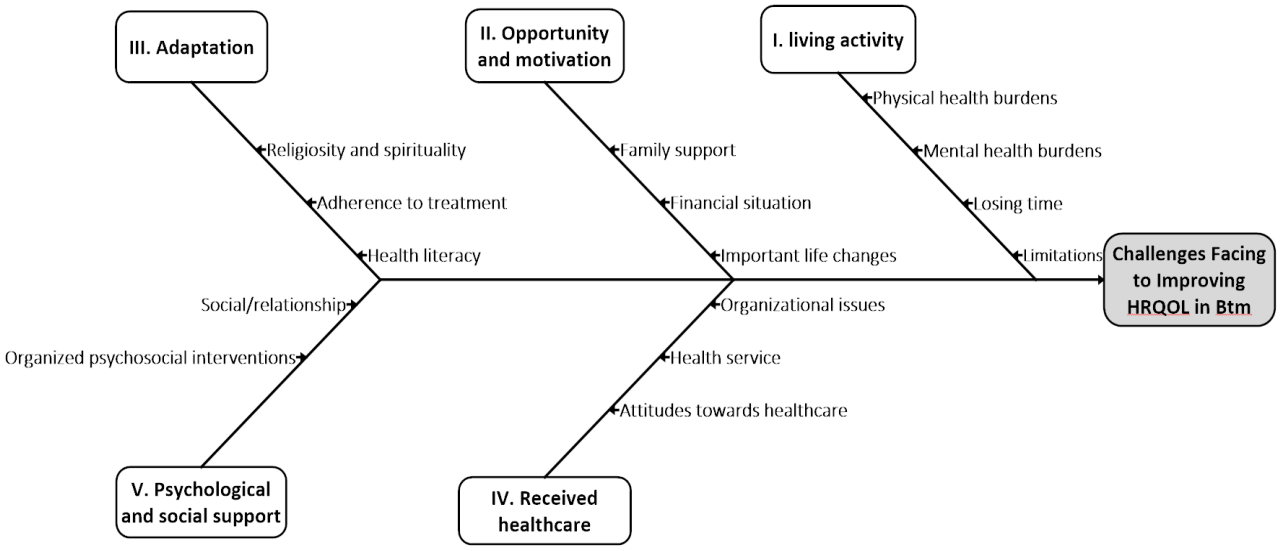

Fig. 1: Fishbone diagram - Facilitators and barriers of improving HRQOL among Iranian β Thalassemia major patients, as reported by the participants

Table-3: Facilitators and barriers of improving HRQOL in Iranian β Thalassemia major patients

Categories	subcategories	Open codes	Barrier or Facilitator
I. living activity	Physical health burdens	Anemia	Barrier
		Fatigue	Barrier
		Shortness of breath	Barrier
		Lack of appetite	Barrier
		Iron overload and its side effects	Barrier
		Infertility	Barrier
	Mental health burdens	Psychological distress	Barrier
		Difficulties with accepting change appearance and face	Barrier
		Lack of autonomy	Barrier
		Ignoring own needs	Barrier
	Losing time	Losing time due to blood transfusion	Barrier
		Losing time due to the preparation of medicines	Barrier
		Losing time during the medical follow-ups or hospitalization	Barrier
	Limitations	Work-inability to continue with the job and occupation	Barrier
		Work-inability to continue with homework and duties	Barrier
II. Opportunity and motivation	Family support	Acceptance of the child's illness	Facilitator
		Refusal to accept a sick child	Barrier
		Emotional support	Facilitator
		Appropriate level of education for parents	Facilitator
		Parental responsibility	Facilitator

Categories	subcategories	Open codes	Barrier or Facilitator
		Distinguish between healthy and sick children	Barrier
		Single child selection	Both of them
		Increasing the age gaps of children	Both of them
		Agreement with the sick child's marriage	Both of them
		Opposition to the marriage of a sick child	Both of them
		Poor family support	Barrier
	Financial situation	Financial strength	Facilitator
		Lower income	Barrier
		More expenses	Barrier
	Important life changes	Getting married	Both of them
		Get pregnant	Both of them
		Having a child/children in life	Both of them
		Acquisition of academic education	Both of them
		Employment and achieving a suitable job	Facilitator
		Bone marrow transplant	Both of them
III. Adaptation	Religiosity and spirituality	Trust in God	Facilitator
		Sincere worship	Facilitator
		Faith in God	Facilitator
		Belief in destiny	Facilitator
		Relying on religious values	Facilitator
	Adherence to treatment	Medical follow-up	Facilitator
		Side-effects	Barrier
		Non-adherence to treatment	Barrier
		Discontinuing current treatment	Barrier
		Seeking more effective medications	Facilitator
		Testing new medication	Facilitator
		Changing iron chelators from injectable to oral	Facilitator
		Development of iron chelators	Facilitator
	Health literacy	Understanding the cause of disease	Facilitator
		Information on medications	Facilitator
IV. Received healthcare	Organizational issues	Conflicting orders from various physicians	Barrier
		Collaboration between different healthcare specialists	Facilitator
		Short visit time by a specialist	Barrier
		Hard access to specialists outside the Thalassemia Clinic	Barrier
		Receiving medication at the treatment site	Facilitator
		Access to a dedicated treatment environment	Facilitator
		Efficient management of the thalassemia center	Facilitator
		Lack of specialist nurses in thalassemia center	Barrier
		Lack of adult hematologists in thalassemia center	Barrier
	Health service	Insufficient supply of original iron chelators	Barrier
		Insufficient supply of packed red cel blood	Barrier
		Lack of life insurance coverage	Barrier
		adequate equipment provided for treatment and screening side effects	Facilitator

Categories	subcategories	Open codes	Barrier or Facilitator
	Attitudes towards healthcare	Lack of trust in healthcare teams	Barrier
		Lack of trust to pharmaceutical companies	Barrier
		Low perceived need to visit healthcare professionals	Barrier
		Unrealistic expectations	Barrier
		Frustration with doctors	Barrier
		Limited skills and expertise of the physicians regarding the patients' behavioral change	Barrier
		Limited skills of healthcare providers	Barrier
		Lack of (interpersonal) skills by healthcare professionals	Barrier
V. Psychological and social support	Social/relationship	Social isolation	Barrier
		Disease concealment due to Social Stigma	Barrier
		Social and Non-governmental organization (NGO)supports	Barrier
		Changes in relationships with family and friends	Barrier
		Changes in work-family balance	Barrier
		New social and family responsibilities	Both of them
		Social context with traditional beliefs	Barrier
	Organized psychosocial interventions	Lack of mental health services for thalassemia major patients in thalassemia center	Facilitator
		Psychosocial interventions for thalassemia major patients	Facilitator
		Psychosocial interventions for parents and family members of thalassemia major patients	Facilitator
		Family Therapy	Facilitator

3-1- Living Activity

All participants confirmed that the physical complications of thalassemia (anemia, fatigue, shortness of breath, lack of appetite, iron overload and its side effects, infertility) have led to physical health burdens and overshadowed the patient's daily life and activities.

"My anemia is like a car running out of gasoline. When I am anemic, I have no energy and I get bored. My bones hurt and I can't do my daily routine. Due to the deposition of iron in my heart, I have heart failure; and the slightest physical activity makes me feel short of breath". (Participant No. 1, a 28-year-old single male patient)

Most patients were worried about the disease and its treatment. They mentioned the changes in their appearance, fear of the worst happening or death panic. Mental health burdens in some patients led to a lack of attention to their needs. They were totally uninterested in life and could not enjoy life, and their daily functioning was also disrupted.

"My illness and its complications have caused me to lose my self-confidence. I feel absurd and I am disappointed with myself and the world in which I live. I reckon that I will never get better and continuing to live is of no value to me". (Participant No. 2, a 21-year-old married female patient)

Almost half of the subjects believed that their lifelong treatment was the main cause of losing time in their lives.

"I have lost too much time in my life due to monthly blood transfusions, regular visits to the specialist and repeated tests. It sounds to me, that the useful days of a thalassemia patient are less than those of healthy people". (Participant No. 3, a 22-year-old married female patient)

52% of the participants considered regular injections of iron chelator, the need for care and treatment and physical complications as limitation for traveling, finding a suitable job, doing daily routine and doing homework.

"Due to my heart disease, I cannot do my favourite activities and sports. Also, I go to school late in the morning and I totally miss my classes on blood transfusion days, and due to night iron injections". (Participant No. 7, a 18-year-old single male patient)

3-2- Opportunity and Motivation

According to the analysis, all participants believed that family support played the most prominent role in the patient's life, and the patients' mothers had become the substantial supporters for thalassemia patients. The patient needed constant family support in the arduous path of life. Responsibility, education level and emotional support of parents were the factors that reduced the complications of thalassemia and played a role in creating life satisfaction in him/her; while lack of cheerfulness in the family due to having an ill member was one of the causes of patients' dissatisfaction with their lives.

"The family is the most important segment for caring and supporting of thalassemia patients. I believe that on time follow ups of dutiful parents can be helpful for treatment of their thalassemia children and may prevent irremediable complications in their adulthood". (Participant No. 20, a 43-year-old married female physician at Thalassemia Clinic)

All participants considered the financial situation as an overarching factor in having a carefree life; besides, they believed that thalassemia imposed a heavy financial burden on the patient and his/her family. Sometimes economic pressure hindered the supply of high quality medication, regular visits to a specialist, doing screening tests and even providing proper nutrition.

"Treating and caring is costly for a thalassemia patient. Recently, the price of original iron chelator medications has increased, and I can no longer afford them. I have not even had an echocardiogram for two years due to my poor financial conditions". (Participant No. 21, a 30-year-old married female patient)

More than half of the participants considered "important life changes" as a factor influencing life. A patient who had experienced a bone marrow transplant, despite describing it as a horrific experience like a death tunnel, was willing to have the transplant again. Having an academic education, having a good job, marriage and having children increased the patient's hope for life, despite all the difficulties and responsibilities they brought about for the patient.

"Getting married and having children were the most important changes in my life. Although my responsibilities have increased, I'm not lonesome anymore and they made me a more hopeful life. Marrying a healthy wife is the greatest chance of my life, and my children are the biggest motivation in my life". (Participant No. 19, a 29-year-old married male patient)

3-3- Adaptation

All participants cited "religiosity and spirituality" as a major source of support. They repeatedly spoke of trust in God.

"I believe that thalassemia is a gift from God, so I endure the pain and suffering. God is just. Verily, with every hardship comes ease". (Participant No. 14, a 33-year-old married male patient)

On the other hand, all participants stated that "adherence to treatment" was vital for a normal life. In their opinion, treatment side effects, lack of follow-up treatment and quitting therapy were the leading obstacles to have a good life. However, switching intravenous to oral iron chelator drugs and development of iron chelator drugs were represented as the most significant facilitators, because they relieved the patient from discomfort of regular night injections.

"Development of iron chelator medications, especially the production of oral medications, has revolutionized the quality of life for thalassemia patients. Production of these drugs is a global approach which is greatly welcomed by patients due to the ease of use". (Participant No. 11, a 48-year-old married male physician at Thalassemia Clinic)

Most of participants considered "health literacy" as an important factor for effective disease management and informed decision making. They believed that increasing the patients' awareness and knowledge would improve their attitude about the disease and increase their capacity for self-care.

"Disease awareness increases health-promoting behaviors in thalassemia patients and has the potential to promote their health and improve their lives, and thus reduce health care costs due to reduced complications". (Participant No. 10, a 38-year-old married female nurse at Thalassemia Clinic)

3-4- Received Healthcare

More than half of the participants stated that the care services, cooperation of the specialists and the management in the thalassemia clinic were appropriate. Some thalassemia patients mentioned the negative impact of conflicting comments from various physicians (especially in comorbidities). Some patients declared that the time to visit a specialist at the Thalassemia Clinic was short and they could not discuss all their problems with the physician in that short time. The most significant challenges in the health care system from patients' point of view were the difficult access to specialists outside the thalassemia clinic, the lack of experienced nurses and the shortage of adult hematologists, which all affected their treatment and life.

"Ever since a dedicated environment for thalassemia patients was established, various specialists have been working together to treat thalassemia patients. Providing care and treatment files and records in this care center has played an effective role in increasing the life expectancy of thalassemia patients". (Participant No. 20, a 43-year-old married female physician at Thalassemia Clinic)

"Unfortunately, adult thalassemia patients are still visited by a pediatrician at the thalassemia care center, and even if a patient becomes pregnant, she will be visited by a pediatrician". (Participant No. 16, a 35-year-old married female patient)

More than half of the participants considered Health Service to be responsible for providing iron depleting drugs and healthy blood. All patients were dissatisfied with the shortage of iron depleting medications, the lack of life insurance coverage for thalassemia patients, and the lack of screening equipment such as MRI; and they wanted a change in national health policy.

"I have not received any good iron chelator medication in three months; every time I try, they say we no longer import medicine. Last month, I had a delay in blood transfusions due to lack of blood. Plus, I have to go to another city for performing annual MRI because they don't perform it here". (Participant No. 15, a 24-year-old single female patient)

All patients' attitudes towards healthcare in the health care system was negative. Reasons for this negative attitude included lack of trust in new drugs and pharmaceutical companies, lack of professional skills, weak interpersonal skills of the clinical staff, caregivers' inability to change patients' behavior, dissapointment with the physician, and physicians' encountering unrealistic patient expectations. Participants stated that if there was a positive attitude towards the healthcare system, the patient's trust in healthcare would be increased and it would have a positive effect on the patient's health and life. Meanwhile, the specialists defined that patients sometimes had unrealistic expectations about diagnosis or treatment that affected their attitudes.

"Some people expect to take a pill and everything will be fine. This is not realistic!" (Participant No. 11, a 48-year-old married male physician at Thalassemia Clinic)

"I was disappointed with the clinic doctors; I do not trust them anymore. I was told that it's not possible for a thalassemia patient to have children, while a number of patients have become pregnant in this city. One of the doctors told me that there was no cure for my heart disease, but after visiting another cardiologist and taking medicine, my heart complications disappeared and now, according to the cardiologist, I do not need heart medicine". (Participant No. 5, a 38-year-old married male patient)

3-5- Psychological and Social Support

All participants believed that supporting organizations such as Thalassemia Association had an impressive effect on improving social relationships. Some patients and their families hid thalassemia for fear of social stigma or certain cultural beliefs. Participants believed that thalassemia exerted considerable pressure on parents and had a negative impact on family relationships. Some patients opined that accepting new responsibilities in the family and community was difficult at first, but over time it became easier and played a key role in improving their quality of life.

"Thalassemia Association helps to increase the presence of thalassemia patients in the community, and on the other hand, has an effective role in countering misbeliefs about thalassemia patients in the community through advertising programs". (Participant No. 12, a 28-year-old married patient's family member)

More than half of the participants mentioned that organized psychosocial interventions were vital for the survival of thalassemia patients. Thalassemia patients are psychologically vulnerable due to therapeutic stress, complications of the disease and having a lifelong illness; hence, psychological interventions and psychiatric counselling have a facilitating role in their lives. On the other hand, the parents of these patients are also under great pressure and sometimes they are not able to deal with their child's illness and they suffer from mental and emotional complication, and are not able to take care of their unhealthy child; psychological counseling programs and family therapy can, thus, help improve this situation.

"Having a child with thalassemia sometimes makes the mothers of these patients very depressed, disrupts family relationships, and reduces the family's

ability to care for the child. In these cases, families are referred to counselling for family therapy". (Participant No. 23, a 43-year-old married patient's family member)

"Counselling interventions and family therapy have played an effective role in improving the mental health of patients and their families. Family's mental health directly affects the mental health of a thalassemia patient and vice versa". (Participant No. 10, a 38-year-old married female nurse at Thalassemia Clinic)

4- DISCUSSION

The present qualitative study investigates the challenges of promoting HRQOL in beta thalassemia major patients with ≥18 years of age, in an Iranian population. After analyzing the data, the challenges were categorized into five major themes, including I. living activity, II. opportunity and motivation, III. adaptation, IV. received healthcare, V. psychological, and social support.

Patients certified that the thalassemia therapy and its complications directly affected their daily performance. Overnight iron chelator injections and frequent blood transfusions prevented them from doing their favorite activities and studying. In terms of general health, they were more susceptible to get any disease than others. Inadequate growth, different physical appearance, uncertain future and expectation of premature death had negative impact on their mental health and they felt desperate in life. Ismail et al. (2006), who studied HRQOL in children with thalassemia in Kuala Lumpur and Malaysia reported that the physical, social, and school performance scores in these children were significantly lower than in healthy children (about 29% to 74%) (18). In these patients, the grief of losing their health and the thought that their lives were different from healthy people caused depression and low self-esteem. In addition, their HRQOL was lower in physical, mental and social dimensions than that of the healthy individuals (7, 19, 20).

In the present study, health care providers believed that the acceptance of the child's illness and parents' responsibility and awareness had a positive impact on patients' HRQOL. A study by Mangione-Smi et al. (2011), illustrated that the right attitude, increased awareness and proper practice of parents led to effective child care, reduced school absenteeism, reduced non-emergency visits to medical centers and an overall increase in child activities in the community. It also resulted in the promotion of HRQOL (21). In the present study, parents gave more freedom to a child with thalassemia than other children due to feeling guilty for the birth of such a child. For thalassemia patients, this parental action showed the importance of such a child to the parents. Another approach of the parents was to make the decision of having just one child or increase the age gap between children, especially the age gap between the thalassemia child and the next child to spend more time for caring and treating a child with thalassemia. According to healthcare professionals, this age gap between children due to more support from parents to the thalassemia child had a positive impact on reducing complications and improving patients' HRQOL.

Patients admitted that they sometimes quit the treatment because they could not afford to pay for medical visits, laboratory services, medications, transfer to medical centers, and hospitalization, which had a negative impact on their lives. According to a study by Emamgholipour et al. (2018), treating each patient with beta thalassemia major costed an average of $ 8,321.8 per year. With a 3% discount rate, the cost of treatment for each patient was estimated at $ 147,098.4, resulting in QALY (Quality-adjusted life year) = 11.8. Therefore, the cost of creating each QALY was $

12,466.0. In their study, by increasing age, QALYs provided by treatment was decreased and the costs were increased (22). In Ho's study in Taiwan, the annual cost of treatment for these patients was $ 7,464.4, which was in line with the findings in Iranian studies (23). In the present study, patients elucidated that due to the family's inability to afford medical expenses, they had to find unsuitable jobs from an early age, despite their physical complications; this resulted in more complications. This finding was consistent with previous studies (7, 24, 25).

In the present study, parents believed that thalassemia patients could not manage their lives independently and had to marry a healthy person; they were opposed to the marriage of two partners who were carriers of thalassemia major. This view of parents had adverse effects on the emotional and mental state of their thalassemia children. A study by Abedi et al. (2014), in Iran showed that parents' opposition and strictness towards marriage of thalassemia patients was one of the significant concerns of patients; and had a negative impact on the psychological dimension of HRQOL in thalassemia patients. However, there was not such a concern in studies of other countries. Thus, given the importance of marriage in Iranian-Islamic culture, it is necessary to change the culture and viewpoints of people in this society; and by changing the view towards the marriage of these patients, this distress should be removed (26).

In the present study, consistent with previous studies (7, 27), thalassemia patients considered their disease as a gift from God (7, 27) and believed that they should strive to keep it. These patients communicate with God by reciting the Holy Qur'an, saying prayer, and meditating; these spiritual practices bring them about self-care increase and anxiety reduction. Spiritual beliefs, in addition to increasing the ability of thalassemia major patients to accept their disease, led to an increase in the responsibility for self-care, which was the basis for adaptation to illness. Parsian and Dunning (2009) suggested a significant and positive relationship among adaptation to illness, reducing complications and spirituality. Patients who scored higher on spirituality had significantly lower hemoglobin A1c levels. According to their findings, spirituality and self-consciousness were helpful in adaptation of patients with chronic diseases to stressful situations; and they should be considered as important aspects in taking care of patients with chronic diseases (28).

In the present study, patients' right attitude and knowledge about treatment and the presence of oral iron chelator drugs caused treatment adherence and reduced complications, which was consistent with previous studies (29, 30). The specialists participating in the present study stated that oral iron chelator drugs, due to their ease of use, led to increased treatment adherence and reduced physical complications that increased the HRQOL; it was consistent with the results of previous studies (7, 25, 26).

Some patients believed that some of the barriers that indirectly reduced HRQOL were receiving conflicting comments from physicians, poor communication between patients and caregivers, time constraints for receiving services, especially short visits by a physician, and distrust of the healthcare system. According to a study by Sedlar et al. (2020), poor communication between caregivers and the patient was an important barrier to the follow-up of the disease complications, since the patient could not discuss all his/her problems with the physician. This led the patient to place unrealistic expectations on treatment. The patient did not follow the treatment according to the treatment protocol and in addition to physical complications, the patient felt hopeless about the treatment

(10). In the present study, some patients complained that their needs were ignored by the health care providers. According to Tao et al.'s study (2020), some health care providers might not be aware of the actual needs of patients with chronic diseases and did not know how to support them in the healthcare setting or community; so, planning and establishing effective communication between the healthcare workers and patients to exchange views and detect patients' problems (31) led to more effective services, reducing patient complications and increasing their HRQOL (32).

In the present study, despite the existence of a specialized thalassemia care center in eastern Iran, patients had to go to another city due to lack of access to MRI for the annual quantifying of iron overload in their body. This transfer to another city was a barrier to perform MRI (Magnetic resonance imaging). According to Waheed (2015), the treatment and screening of thalassemia symptoms in a city other than the patient's hometown, in addition to the financial burden, imposed difficult travel conditions on them and if the patient did not go to this travel, there would be a possibility of increased complications (7). Therefore, providing screening facilities to investigate iron deposition in the patient's organs in thalassemia care centers should be one of the priorities of the healthcare system in countries affected by thalassemia for providing convenient conditions in the treatment of patients (3, 30).

The findings of the present study exhibited that the parents' concern for the ill child and constant monitoring of the treatment brought about a stressful situation with frustration for the parents, which interrupted the caregiving and indirectly had a negative impact on the HRQOL of the thalassemia patient. This finding was consistent with Ishfaq et al.'s study (2015) (33). Some patients stated that due to their illness, they avoided social activities and discussing their illness with relatives, friends and neighbors because they felt that they were not understood by others, and this led to their social isolation. This was also in line with previous studies (7, 32). Hence, psychosocial support and family therapy are considered as essential aspects of care management to promote HRQOL in chronic patients (34, 35). Promoting community knowledge and planning for the presence of these patients in the community should be included in the health care policies for these patients (1, 24).

5- STRENGTHS AND LIMITATIONS

This study was conducted by an interprofessional team through selecting participants of maximum diversity; and it provided the opportunity to integrate views and opinions. The present population includes patients, family members and caregivers from eastern Iran, so it may not represent all thalassemia patients in Iran. Therefore, transferability of findings to other cultures and contexts needs further studies.

6- CONCLUSIONS

By the development of treatment and iron chelator drugs, governments are facing an increase in the age of thalassemia major patients. Increasing the age of these patients is associated with increased physical, psychological, social complications and their treatment costs, which lead to a decrease in their HRQOL. Thus, in parallel with age increase, it is necessary to plan for their presence in society and peculiar measures should be taken for employment, education and marriage of patients with thalassemia major. The treatment of these patients is reaching the cost-effectiveness threshold as their life expectancy increases, so policy-making for screening the complications is vital. In screening, it is not just enough to provide a medical

environment without supplying appropriate equipment. Moreover, the availability of laboratories or MRI equipment to measure iron deposition near thalassemia care centers should be one of the priorities of the healthcare system of countries affected by thalassemia in order to provide convenient treatment conditions for patients. Sometimes patients' unrealistic expectations about thalassemia treatment are barriers for improving their health. In this regard, informing them about treatment and illness leads to a right attitude and lowers their unrealistic expectations. The physical limitations and appearance changes of these patients are the reasons of their not being accepted by the society; and in many families, these patients are deprived of marriage. Therefore, by the accurate culture making in society and changing the views of people and parents, worries of these patients can be calmed.

7- CONFLICT OF INTEREST

The authors declare no conflicts of interest.

8- REFERENCES

1. Arian M, Mirmohammadkhani M, Ghorbani R, Soleimani M. Health-related quality of life (HRQoL) in beta-thalassemia major (beta-TM) patients assessed by 36-item short form health survey (SF-36): a meta-analysis. Quality of life research : an international journal of quality of life aspects of treatment, care and rehabilitation. 2019;28(2):321-34.

2. Razzazan N, Ravanipour M, Jahanpour F, Zarei AR, Motamed N, Hosseini S. Investigating Challenges Facing Self-Management Empowerment Education in Adolescents and Youths with Major Thalassemia. Jundishapur J Chronic Dis Care. 2014;3(4):e23366.

3. Arian M. The effect of designed walking program with holistic nursing approach on serum ferritin and heart problems of Thalassemia Major patients. Tehran: Tarbiat Modares University; 2013.

4. Cappellini M, Cohen A, Eleftheriou A, Piga A, Porter J, Taher A. Guidelines for the Clinical Management of Thalassaemia [Internet]: Thalassaemia International Federation TIF; 2008.; 2014.

5. Ibrahim H, Hassan A, George E, Goh A. Management of Transfusion Dependent Thalassaemia. Malaysia: Ministry of Health and Academy of Medicine. 2009:50-2.

6. Vizza J. A whole new world: the experiences of adolescents with beta-thalassemia major as they transition to adult care. Ontario University of Ontario Institute of Technology 2018.

7. Waheed F. Thalassaemia prevention in Maldives: effectiveness of primary, secondary and tertiary prevention interventions. Western Australia The University of Western Australia; 2015.

8. Biswas B, Naskar NN, Basu K, Dasgupta A, Basu R, Paul B. Care-Related Quality of Life of Caregivers of Beta-Thalassemia Major Children: An Epidemiological Study in Eastern India. J Epidemiol Glob Health. 2020;10(2):168-77.

9. Willison KD, Andrews GJ. The potential of public health to enhance chronic disease management. Public health. 2005;119(12):1130-2.

10. Sedlar N, Lainscak M, Farkas J. Living with Chronic Heart Failure: Exploring Patient, Informal Caregiver, and Healthcare Professional Perceptions. International journal of environmental research and public health. 2020;17(8):2666.

11. Ford S, Schofield T, Hope T. Barriers to the evidence-based patient choice (EBPC) consultation. Patient Educ Couns. 2002;47(2):179-85.

12. Kennedy BM, Jaligam V, Conish BK, Johnson WD, Melancon B, Katzmarzyk PT. Exploring Patient, Caregiver, and Healthcare Provider Perceptions of Caring for Patients With Heart Failure: What Are the Implications? Ochsner J. 2017;17(1):93-102.

13. Elo S, Kyngäs H. The qualitative content analysis process. Journal of advanced nursing. 2008;62(1):107-15.

14. Vaismoradi M, Jones J, Turunen H, Snelgrove S. Theme development in qualitative content analysis and thematic analysis. Journal of Nursing Education and Practice. 2016;6(5):100-10.

15. Vaismoradi M, Turunen H, Bondas T. Content analysis and thematic analysis: Implications for conducting a qualitative descriptive study. Nursing & health sciences. 2013;15(3):398-405.

16. Guba EG, Lincoln YS. Fourth generation evaluation: Sage; 1989.

17. Assarroudi A, Heshmati Nabavi F, Armat MR, Ebadi A, Vaismoradi M. Directed qualitative content analysis: the description and elaboration of its underpinning methods and data analysis process. Journal of Research in Nursing. 2018;23(1):42-55.

18. Ismail A, Campbell MJ, Ibrahim HM, Jones GL. Health related quality of life in Malaysian children with thalassaemia. Health and quality of life outcomes. 2006;4(1):39.

19. Mazzone L, Battaglia L, Andreozzi F, Romeo MA, Mazzone D. Emotional impact in beta-thalassaemia major children following cognitive-behavioural family therapy and quality of life of caregiving mothers. Clinical practice and epidemiology in mental health : CP & EMH. 2009;5:5.

20. Roudbari M, Soltani-Rad M, Roudbari S. The survival analysis of beta thalassemia major patients in South East of Iran. Saudi medical journal. 2008;29(7):1031-5.

21. Mangione-Smith R, Schiff J, Dougherty D. Identifying children's health care quality measures for Medicaid and CHIP: an evidence-informed, publicly transparent expert process. Academic pediatrics. 2011;11(3 Suppl):S11-21.

22. Emamgholipour S, Ahmadi B, Rajabi AH, Azarkeivan A, Ebrahimi M, Esmaeilzadeh F. Cost-utility of treatment of the patients with Thalassemia Major in Iran. The Scientific Journal of Iranian Blood Transfusion Organization. 2018;15(4):257-64.

23. Ho WL, Lin KH, Wang JD, Hwang JS, Chung CW, Lin DT, et al. Financial burden of national health insurance for treating patients with transfusion-dependent thalassemia in Taiwan. Bone marrow transplantation. 2006;37(6):569-74.

24. Heidari H, Ahmadi A. Change in the Health System Structure is a Way to Support Parents Having Children with Thalassemia: A Qualitative Study. Jundishapur J Chronic Dis Care. 2019;8(3):e86095.

25. Hisam A, Khan NUS, Tariq NA, Irfan H, Arif B, Noor M. Perceived stress and monetary burden among thalassemia patients and their caregivers. Pak J Med Sci. 2018;34(4):901-6.

26. Abedi H, Abbaszadeh A, Kazemi M, Pouraboli B. Silent Screams: Experiences of Caregiver Suffering by Parents of Children with Thalassemia: A Qualitative Study. ournal of Qualitative Research in Health Sciences,. 2014;3(3):281 - 91.

27. Yousefian Z, Ahadi H, Keraskian Mojmanari A. The effectiveness of spirituality therapy on psychological distress and resilience of thalassemia patients

Journal of Psychological Science. 2020;18(84):2285-93.

28. Parsian N, Dunning T. Spirituality and coping in young adults with diabetes: a cross-sectional study. European Diabetes Nursing. 2009;6(3):100-4.

29. Valliammal Shanmugam, Ramachandra. Self-Care Deficits in Adolescents with Thalassemia: Qualitative study. Int J Adv Nur Management. 2014;2(2):55-60.

30. Zeydi AE, Moonaghi HK, Heydari A. Exploring Iranian β-Thalassemia major patients' perception of barriers and facilitators of adherence to treatment: A qualitative study. Electronic Physician. 2017;9(12):6102.

31. Tao WW, Tao XM, Wang Y, Bi SH. Psycho-social and educational interventions for enhancing adherence to dialysis in adults with end-stage renal disease: A meta-analysis. J Clin Nurs. 2020;29(15-16):2834-48.

32. Pouraboli B, Abedi H, Abbaszadeh A, Kazemi M. The Burden of Care: Experiences of Parents of Children with Thalassemia. J Nurs Care. 2017;6:389.

33. Ishfaq K, Shabbir M, Naeem SB, Hussain S. Impact of Thalassemia Major on Patients' Families in Pakistan The Professional Medical Journal. 2015;22(05):582-9.

34. Ebrahimi H, Hasankhani H, Namdar H, Khodadadi E, Fooladi M. Dealing with Chronic Illness: Experiences of Iranian Families of Persons with Multiple Sclerosis—A Qualitative Study. Multiple sclerosis international. 2017;2017.

35. Golics CJ, Basra MKA, Finlay AY, Salek S. The impact of disease on family members: a critical aspect of medical care. Journal of the Royal Society of Medicine. 2013;106(10):399-407.