Iran J Public Health, Vol. 51, No.1, Jan 2022, pp.12-18

Review Article

Thalassemia Major and Associated Psychosocial Problems: A Narrative Review

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(Received 11 Sep 2021; accepted 07 Nov 2021)

Abstract

Thalassemia is an inherited disease that causes the production of damaged hemoglobin chains. Patients are diagnosed with thalassemia major due to major clinical signs and deep anemia. This study aimed to examine the major thalassemia and psychosocial aspect of it, which is such an important issue, to serve as a roadmap for better handling these patients and to contribute to the literature. The method used in this study was narrative review. A literature review was conducted by searching the materials published in databases including Web of Science, PubMed, Scopus, and Google Scholar search engine from 2001 to 2020. Besides WHO website was searched. Thalassemia major damages the heart, liver, lungs and endocrine organs due to anemia and iron accumulation. In addition, the patient may experience mental and social problems due to the congenital nature of the disease and its lifelong duration. The psychosocial problems and treatment burdens of thalassemia patients are very high. There are many studies about the prevalence and physical consequences of thalassemia. However, there are not enough articles and researches describing the psychosocial effects of thalassemia on patients and what can be done about these effects. For this reason, this paper focuses on the process of thalassemia and the psychosocial problems it creates to contribute to the literature and to be a roadmap for better handling these patients.

Keywords: Thalassemia major; Blood disease; Psychosocial problem; Psychosocial care

Introduction

Thalassemia is an autosomal recessive blood disease characterized by anemia that develops because of damaged synthesis of one or more of the hemoglobin chains. When thalassemia is classified considering the clinical situation; patients with very little or no anemia despite having abnormal erythrocyte structure are classified as thalassemia minor (carrier, heterozygous), patients whose anemia does not require regular transfusion are classified as thalassemia intermedia (pa-

tient, homozygous), patients with major clinical findings and deep anemia are classified as thalassemia major (patient, homozygous) (1,2). Thalassemia is a major health problem that threatens 72% of 229 countries around the world. 5.2% of the world population, 7% of pregnant women, and 1% of couples are at risk of thalassemia. According to WHO, thalassemia carriage is 5% in the world (3). About 17% of babies born every year carry the thalassemia gene (4). Thalassemia has different distributions in many countries and different parts of the countries. Among Mediterranean countries, the density has increased in Europe and the United States of America with the migrations from the Middle East and the Far East. The density is also increasing along the coast, which includes the Mediterranean area, Greece, Italy, Cyprus, Malta, Spain, and western and southern coasts of the Republic of Turkey and in the far eastern countries (5). Thalassemia major is one of the common causes of anemia in the Mediterranean countries. Thalassemia major is an important public health problem that is inherited from parents to children, can be prevented by screening programs, progresses severely when not treated, negatively affects the quality of life and shortens life expectancy (6). Therefore, thalassemia major will be the focus of this article.

Thalassemia major shows clinical indications between 6 months and 2 yr of age. Damage occurs in the heart, liver, lungs and endocrine organs due to anemia and iron overload (7). Patients with thalassemia experience growth retardation, shape changes in the face, head and teeth due to bone deformities, cardiac and hepatic complications, delayed puberty, diabetes mellitus, hypothyroidism and hypoparathyroidism. The treatment of thalassemia major includes erythrocyte transfusion, iron chelation therapy, surgical treatment such as splenectomy, as well as follow-up and treatment of related complications with psychological support, and stem cell transplantation (6,8,9).

New developments in medical technology and treatments applied in centers with rich resources prolong life in thalassemia. Developed and developing countries include "thalassemia prevention and treatment programs" in their national health policies. In this context, Mediterranean countries have been successful in fighting against this disease by performing screening tests for couples who want to have a baby or before marriage. Pre-marital screening is preferred by countries because it is easy to apply and economical. The purpose of the scan is to identify risky couples and to enable them to have healthy children by providing genetic counseling. The screening method is effective in preventing the birth of children with thalassemia (2,10).

Like all chronic diseases, thalassemia major also affects patients' lives. It is a disease that has negative physical, mental and social consequences (11,12). Thanks to the developments in thalassemia major treatment, the implementation of transfusion protocols has shown an increase in the life span and quality of life of patients (6,13). However, complications due to treatment may occur because of low reliability of blood and blood products, inadequate blood transfusion therapy, chronic iron deposition, or inadequate use of chelation therapy. Patients experience musculoskeletal disorders, bone deformations, and osteoporosis problems. Their quality of life decreases due to severe pain decreased functional capacity and osteoporosis.

Thalassemia major is a disease that negatively affects the patients psychosocially. There are physical malformations, growth retardation, hepatosplenomegaly, yellow skin color and characteristic facial appearance with prominent lines like the disease. This body image negatively affects the self-esteem and self-confidence of individuals and causes them to feel different from other individuals. Therefore, patients take a passive role in social environments by isolating themselves from society, so their quality of life and mental states are negatively affected (11,14). The individual's physical health, psychological condition, level of independence, social relations, personal beliefs and the quality of life, including relationships with the environment, are negatively affected by the chronicity of thalassemia, the need for transfusion at regular intervals throughout life, and the related complications (6,15). Frequent

hospitalization due to transfusion, staying away from the family, activity limitations and pain, side effects of iron chelation therapy, and fear of death can also lead to anxiety, hopelessness and depression. This also increases the negative impact of people and the cost to the country. The prevalence of psychosocial maladjustment was 80% in patients with thalassemia major. Patients may experience many emotional problems, anxiety, and behavioral problems in various periods of their life. These are frustration, grief, hostility, anxiety, hopelessness, inability to cope with anxiety, fear of death, lack of confidence, somatization, decreased self-esteem, isolation, irritability, helplessness, feeling of lovelessness and lack of self-respect (16-19).

Thalassemia is an important public health problem for the Mediterranean countries. Thalassemia patients have many psychosocial problems due to the reasons mentioned above. The sources are written are very limited in this context. Even if the patients are followed physically, the psychosocial studies are insufficient and cause the patients to have problems. For this reason, this study aimed to examine the major thalassemia and psychosocial aspect of it, which is such an important issue, to serve as a roadmap for better handling these patients and to contribute to the literature.

Psychosocial Problems of Patients with Thalassemia Major

Chronic diseases are persistent long-term diseases, which do not give individuals much chance to shape their future fully. Many chronic diseases require important adjustments in an individual's life. The periods of exacerbation and remission of the disease and changes in health can strain the coping mechanism of the patient and their family. In addition, changes in the routine such as taking the role of caregiver, changes in sexual life, social relations and material life affect the psychosocial response to chronic diseases. Chronic diseases include processes that require adaptation. Poor adaptation can cause depression, denial, decreased self-esteem, addiction, and disharmony (11,20,21). Patients struggling with thalassemia, which is a chronic disease, experience physical and mental trauma. Psychological and social problems are increasing due to the positive developments in the course of the disease and the increasing number of patients reaching adulthood. Like people with other chronic diseases, patients with thalassemia major have to cope with many conditions. While trying to keep the course of the disease under control, mental health is generally ignored (16,17).

Although the difficulties experienced by patients with thalassemia decrease with medical treatment, psychosocial problems are of primary importance in these patients. Common problems are the chronicity of thalassemia major disease, the need for blood transfusion at regular intervals throughout life, body image disorders, being evaluated differently from peers due to the delay in growth and development, high treatment costs, difficulties in recruitment, the need for regular leave to continue treatment throughout their working life and education. All of this requires a constant effort to overcome serious health problems throughout the life of patients with thalassemia major. Therefore, psychosocial problems may be the most important problems for these patients (16,17).

Problems such as social disintegration, job problems, sexual problems, and the desire to establish a family also cause an individual's anxiety to increase. Severe anemia cause patients to feel weak. Transfusions performed to keep the hemoglobin amount at an optimal level help reduce these symptoms and anxiety seen in patients. While regular monthly blood transfusions help patients feel better, they also bring extra problems such as blood-borne virus diseases and high iron levels. This situation causes psychological reactions for treatment in patients (16). Continuous chelation therapy becomes an important burden in the daily lives of patients and negatively affects their psychological and social lives.

Adaptation to disease is a process of getting used to internal and external changes; it includes effective coping and reconciliation. Although psycho-

social adjustment to the disease is affected by all factors related to the disease and treatment, it may affect the course of the disease positively or negatively. Good psychosocial adaptation may enable the patient to cope better with the effects of the disease, to have fewer complications and attacks, and to have less problems in social and family environments. It can increase the patient's sense of control over their disease. In addition, the patient's compliance with the disease increases with the ability of the patient to accept the losses caused by the disease and to use their existing potential for their purposes, to show the flexibility to change their goals when necessary (16,22,23).

Psychological support and treatment for psychosocial problems are not given in the many treatment centers. Biological, psychological and social well-being and patient satisfaction will increase with the biopsychosocial care model in the treatment of patients with thalassemia. In this context, it is also important to consider the psychosocial problems experienced by patients with thalassemia in detail.

Body Image and Self-Esteem in Thalassemia Major Patients

An individual has a holistic structure that needs to be handled in terms of physical and mental structure, social and intellectual aspects. The concept of body includes all perception and knowledge of the individual regarding the external appearance and internal structure of the body. Body image is shaped by all of the individual's thoughts about their personality, values, and relationships with other people. When the individual perceives and evaluates their own body, it plays an important role in determining their body, self-esteem and confidence. Body image consists of physical, psychological and social experiences. Individual who thinks positively about their body image shows a positive attitude, and it means that they tend towards positive personality and self-confidence. Self-esteem is the state of appreciation arising from a person's approval of the self that they reach after evaluating themself. Every person has a concept of self they want to reach, and all efforts of a person throughout their life are aimed at developing the self that they miss and associate with themselves. Chronic diseases can affect self-esteem of patients, as the disease may cause deterioration of body shape and inability to control their own body (24).

Thalassemia major is closely related to psychological conditions such as changes in body image and self-esteem due to bone deformities and growth retardation, and this relationship is clinically important (25,26). Physical deficiency, deformity and physical diseases in chronic diseases reduce self-esteem by distorting body image and creating feelings of inadequacy. Self-esteem and body image levels affect resistance to psychological and physiological diseases; individuals with low self-esteem and body image refuse positive feedback and do not cooperate in treatment. Individuals with low self-esteem and body image may become depressed (27). Patients experience a lack of self-confidence due to both the fear and anxiety caused by thalassemia, which is a chronic disease, and physical changes and weaknesses. Lack of self-esteem is important since it may cause patients to damage their social relationships and stop fighting against the disease.

Life Time and Quality of Patients with Thalassemia Major

The lifetime and life quality of patients with thalassemia major differ depending on whether the recommended conventional treatment protocols are applied or not. Significant improvements have been made in the life span and life quality of patients in the last 10-20 years, thanks to innovations in diagnosis and treatment in the field of thalassemia (28,29). The life expectancy is prolonged because of preventing the complications related to iron load with effective chelation therapies. The discovery and widespread use of Desferal used in chelation therapy constituted an important milestone in the treatment of thalassemia. The biggest problem is the complications that develop because of patients not using chelator agents at recommended doses, even in patients with thalassemia in countries with good treatment opportunities. This directly affects their

quality of life. Therefore, healthcare professionals and their families dealing with thalassemia treatment should not only educate patients but also guide them in the application of effective chelation therapies (15,30).

Patients with thalassemia have been treated within the framework of the protocol recommended by the Thalassemia Federation since the mid-1980s in the Mediterranean countries. Following regular chelation treatments with Desferal, positive improvements were observed in the life span and life quality of the patients (9,31,32). Significant improvements have been recorded in the quality of life and life span of patients with thalassemia in the following years because of more accurate diagnosis and treatment of cardiological and hepatic iron burden with adequate and safer blood transfusions, newly released oral chelators, endocrinological replacement therapies, and non-invasive MRI methods. Patients with thalassemia major have begun to look to the future more confidently due to advances in treatment, and have been encouraged to lead a normal life, to do sports and participate in cultural activities, education and working life. Many patients continued their education in high schools and universities, started to work, got married, and had a family and children (32). However, this does not mean that there is no need for a psychosocial service. The service requirement dimension may change continuously according to the conditions of the day in this area, the development process and sociocultural characteristics of the people.

Despite the effective conventional practices, adequate and reliable blood transfusions and chelation treatments initiated since the second half of the 1980s, patients lost their lives due to infections, hepatic diseases, various reasons, and especially sudden heart failure. The first significant improvement was detected in the life expectancy and life quality of the patients after the recommended conventional treatment practices, and the average life expectancy was prolonged (15,32).

Patients with thalassemia who reach adulthood begin to manage their lives themselves and start to have anxiety about education, work and family life. During this period, many patients experience a great transformation and realize that they can live longer and have a better quality of life with good treatments. Patients are more willing to apply the recommended treatments. The most important factor underlying the social and psychological problems of patients with thalassemia is their anxiety about their desire to live a long and quality life. The fears and worries of patients about their lives can be relieved largely with the efforts of specialist doctors in thalassemia centers with all kinds of infrastructures. Informing patients and their families about the causes and consequences of the disease and convincing them about the treatments to be applied should be among the most important duties of the physician treating thalassemia. Once patients and their families are relieved about treatments, and adequate medical support is provided, things get easier. As a result of the proper application of the recommended treatments for thalassemia patients, their lifespan and life quality are improving, and the level of fear and anxiety decreases (15,33).

Conclusion

Efforts to increase the life span and life quality of existing patients with thalassemia are still continuing and effective iron chelation with adequate and safe blood transfusions continues. Thanks to the meaningful results achieved in the quality and duration of life with continued treatments, patients and their families look to the future more confidently. When the thalassemia process is examined, patients with thalassemia seem to have adapted to living with a chronic disease, but the psychosocial problems and treatment burdens they experience are at a level considered. Therefore, treatment and care for psychosocial problems should be emphasized. The psychosocial care model should be included in the follow-up of the patients. In addition, the cooperation of the patient, family and healthcare professionals is of great importance to improve the well-being

and positive coping mechanisms of individuals with thalassemia.

Ethical considerations

Ethical issues (Including plagiarism, informed consent, misconduct, data fabrication and/or falsification, double publication and/or submission, redundancy, etc.) have been completely observed by the authors.

Conflict of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

References

1. Origa R (2017). β-Thalassemia. *Genet Med*, 19 (6): 609-19.
2. Prathyusha K, Venkataswamy M, Sridivya Goud et al (2019). Thalassemia: A blood disorder, its cause, prevention and management. *Research Journal of Pharmaceutical Dosage Forms and Technology*, 11: 975-4377.
3. WHO (2006). Thalassemia and other haemoglobinapathies: report by the secretariat. https://apps.who.int/iris/handle/10665/21519
4. Modell B, Darlison M (2008). Global epidemiology of haemoglobin disorders and derived service indicators. *Bull World Health Organ*, 86 (6): 480-7.
5. Aydınok Y, Oymak Y, Atabay B et al (2018). A national registry of thalassemia in Turkey: Demographic and disease characteristics of patients, achievements, and challenges in prevention. *Turk J Haematol*, 35 (1): 12–18.
6. Yeşilipek A (2014). Hematopoetic stem cell transplantation in patients with beta thalassemia major, *Archive and Survey Journal*, 23 (1): 49-59.
7. Taher AT, Weatherall DJ, Capellini MD (2018). Thalassaemia. *Lancet*, 391: 155-67.
8. Sharma R, Seth A, Chandra J, et al (2016). Endocrinopathies in adolescents with thalassaemia major receiving oral iron chelation therapy. *Paediatr Int Child Health*, 36: 22-27.
9. Kleanthous M (2019). Screening and prevention of thalassaemia in Cyprus. *Hemoglobin*, 43 (6): 331.
10. Canatan D (2014). Status of thalassemia and hemoglobinopathies in world and Turkey. *Turkey Journal of Clinical Hematology*, (1): 1-4.
11. Öz F (2015). Psychosocial nursing in cancer. *Türkiye Klinikleri*, 1 (2): 46-52.
12. Öz HS, Öz F (2020). A psychoeducation program for stress management and psychosocial problems in multiple sclerosis. *Niger J Clin Pract*, 23: 1598-1606.
13. Ay Kaatsız MA, Öz F (2020). The effectiveness of psychoeducation given to mothers of children with cancer. *J Psychiatric Nurs*, 11 (2): 129-140.
14. Maden A, Hortu H, Üzüm Ö, et al (2018). Evaluation of anxiety and depression levels of adolescents with thalassemia trait. *İzmir Kâtip Çelebi Üniversitesi Sağlık Bilimleri Fakültesi Dergisi*, 5 (3): 277-280.
15. Anwar K, Waqar S (2018). Psychopathological tendencies and quality of life among patients with thalassemia majör. *Rawal Medical Journal*, 43 (1): 32-38.
16. Elzaree F, Shehata M, Wakell M, et al (2018). Adaptive functioning and psychosocial problems in children with beta thalassemia majör. *Open Access Maced J Med Sci,* 6 (12): 2337-2341.
17. Vosper J, Evangeli M, Porter JB, et al (2018). Psychological factors associated with episodic chelation adherence in Thalassemia. *Hemoglobin*, 42 (1): 30-36.
18. Behdani F, Badiee Z, Hebrani P, et al (2015). Psychological aspects in children and adolescents with major thalassemia: A case-control study. *Iran J Pediatr*, 25 (3): e322.
19. Naderi M, Hormozi MR, Ashrafi M, et al (2012). Evaluation of mental health and related factors among patients with beta-thalassemia major in South East of Iran. *Iran J Psychiatry*, 7 (1): 47-51.
20. Patel P, Beamish P, da Silva TL, et al (2019). Examining depression and quality of life in patients with thalassemia in Sri Lanka. *Int J Non-Commun Dis*, 4: 27-33.
21. Öz F (2001). Uncertainty in illness experience. *Turk J Psychiatry*, 12 (1): 61-68.
22. Ishfaq K, Diah NM, Ali J, et al (2018). Psychosocial problems faced by thalassemia major

patients of district multan. *Pak Pediatr J*, 42 (1): 22-26.
23. Öz F, Demiralp M (2014). For psychosocial nursing general patient care. Academician Medical Press, Ankara, Turkey.
24. Grift TC, Cohen-Kettenis T, Vries AC (2018). Body image and self-esteem in disorders of sex development: A European multicenter study. *Health Psychol*, 37 (4): 334–343.
25. Kelleci M, Doğan S (2001). The examination of nurses' approaches to the individuals who had depression associated with other physical diseases. *Alpha Psychiatry*, 2: 161-68.
26. Punriddum J, Sanasuttipun W, Sangperm P (2018). Factors related to body image of adolescents with thalassemia. *J Nurs Sci*, 36 (1): 57-72.
27. Jorge RT, Brumini C, Jones A, et al (2010). Body image in patients with rheumatoid arthritis. *Mod Rheumatol*, 20 (5): 491-5.
28. Telfer P, Constantinidou G, Andreou P, et al (2005). Quality of life in thalassemia. *Ann N Y Acad Sci,* 1054: 273-82.
29. Ağaoğlu L (2010). Life in thalassemia. *Türkiye Klinikleri,* 3 (1): 9-13.
30. Taheri P, Nooryan K, Karimi Z, et al (2020). Effect of individual psychotherapy with a focus on self-efficacy on quality of life in patients with thalassemia major: A clinical trial. *J Clinic Care Skill*, 1 (2): 49-54.
31. Telfer P, Coen PG, Christou S, et al (2006). Survival of medically treated thalassemia patients in Cyprus. Trends and risk factors over the period 1980-2004. *Haematologica*, 91 (9): 1187-92.
32. Bozkurt G, Baysal E. *Thalassemia syndromes: Thalassemia prevention program in Cyprus and its results.* İstanbul: Ateş Press; 2019.
33. Ceylan S, Çetinkaya B, Karabudak S et al (2018). Examining the factors affecting quality of life of children and adolescents with Beta-Thalassemia. *Journal of Dr Behcet Uz Children s Hospital,* 8 (1): 15-22.