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REVIEW

A Hematologic Disease in the Turkish Republic of Northern Cyprus: Thalassemia Major

Kuzey Kıbrıs Türk Cumhuriyeti'nde Bir Hematolojik Hastalık: Talasemi Majör

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Abstract

Thalassemia is an inherited hematologic disease in which the body makes an abnormal form of hemoglobin. Patients are diagnosed with thalassemia major due to severe anemia and clinical symptoms. Thalassemia major is a common disease in the Turkish Republic of Northern Cyprus (TRNC) and other Mediterranean countries. Thalassemia major is a chronic and severe health problem. Thalassemia major damages organs due to intense iron accumulation and deep anemia. It negatively affects the quality of life and shortens life. Also, due to chronic disease, patients experience psychological and social problems. In this review, we present thalassemia major in the TRNC. The rate of patients has decreased with prevention programs and no sick baby has been born after 2001 in the TRNC. However, existing thalassemia patients have many physical and psychosocial problems and the treatment processes the patients are challenging. Since TRNC is an island that receives immigration, it has a sociological texture that includes different cultures, which can increase the dimensions of this problem. Therefore, this review focuses on the thalassemia process and the psychosocial problems caused by the disease in the TRNC.

Keywords: Cyprus, thalassemia major, hematologic disease, patient, psychosocial care

Öz

Talasemi, vücudun anormal bir hemoglobin formu oluşturduğu kalıtsal bir hematolojik hastalıktır. Hastalara şiddetli anemi ve klinik semptomlar nedeniyle talasemi majör tanısı konur. Talasemi majör, Kuzey Kıbrıs Türk Cumhuriyeti (KKTC) ve diğer Akdeniz ülkelerinde yaygın olarak görülen bir hastalıktır. Talasemi majör, kronik ve ciddi bir sağlık sorunudur. Talasemi majör, yoğun demir birikimi ve derin anemi nedeniyle organlara zarar verir. Yaşam kalitesini olumsuz etkiler ve ömrü kısaltır. Ayrıca kronik hastalık nedeniyle hastalar psikolojik ve sosyal sorunlar yaşamaktadır. Bu derlemede KKTC'deki talasemi majörü sunuyoruz. Önleme programları ile hasta oranı düşmüş ve 2001 yılından sonra KKTC'de hasta bebek dünyaya gelmemiştir. Ancak mevcut talasemi hastalarının birçok fiziksel ve psikososyal sorunu bulunmakta ve tedavi süreçleri hastaları zorlamaktadır. KKTC göç alan bir ada olması nedeniyle farklı kültürleri içinde barındıran sosyolojik bir dokuya sahip olması bu sorunun boyutlarını artırabilmektedir. Bu nedenle bu derleme, KKTC'de talasemi sürecine ve hastalığın yol açtığı psikososyal sorunlara odaklanmaktadır.

Anahtar Kelimeler: Kıbrıs, talasemi majör, hematolojik hastalık, hasta, psikososyal bakım

Introduction

Thalassemia is a group of disorders that prevent the body from producing a sufficient quantity of quality blood. In thalassemia, patients with very little or no anemia despite having abnormal erythrocyte structure are classified as thalassemia minor, patients whose anemia does not require regular transfusion are classified as thalassemia intermedia, patients with major clinical symptoms and anemia are classified as thalassemia major (1).

According to a report published by the World Health Organization (2006), 5.2% of individuals in the world are carriers of thalassemia. In the Turkish Republic of Northern

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Cyprus, the rate of thalassemia carrier is 17% (2). Thalassemia major is one of the common causes of anemia in the Turkish Republic of Northern Cyprus (TRNC). Thalassemia major is a hematologic problem that is inherited from parents to children, can be prevented by genetic screening programs, progresses severely when not treated (3,4). The diagnosis of thalassemia major is made between the ages of 6 months and 2 years with clinical symptoms. Damage occurs in the liver, lungs, heart and endocrine organs due to anemia and iron overload (5). Patients experience growth retardation, shape changes in the face, jaundice, head and teeth due to bone deformities, diabetes mellitus, cardiac and hepatic complications, delayed puberty, infection, osteoporosis, hypothyroidism and hypoparathyroidism. The treatment of thalassemia major includes blood transfusion, iron chelation therapy, splenectomy, psychological care, and stem cell transplantation (4,6).

Mediterranean countries include "thalassemia prevention and treatment programs" in their health policies. In this context, the TRNC conducted screening tests for couples who wanted to get married and received positive results and were successful in combating this disease. The purpose of the scan is to identify risky couples and to enable them to have healthy children by providing genetic counseling. Published reports have shown that genetic screening is effective in preventing birth of children with thalassemia major (7). TRNC started programs to fight against thalassemia in the 1980s and their practices still actively continue. It was reported that the last birth of a baby with thalassemia occurred in 2001 (8). This is important in terms of demonstrating how useful and valuable these preventive programs implemented on a country basis, are in reducing and preventing thalassemia.

Chronic diseases create compelling effects on patients' lives. Thalassemia major, is a chronic disease, affects the lives of patients and causes physical, social and psychological problems (9,10). There are physical deformations, jaundice, growth retardation, hepatosplenomegaly and characteristic facial appearance with prominent lines in the symptoms of the disease. These symptoms, body image, negatively affects the self-esteem and self-confidence of individuals. Therefore, patient's quality of life and psychosocial states are negatively affected (4,11-13). The individual's physical health, psychological condition, social relations, personal beliefs and the quality of life, including relationships with the people, are negatively affected by the chronicity of thalassemia, the need for blood transfusion at regular intervals throughout life, and the related complications (10,13,14). 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, stress, hopelessness and depression (15,16). The studies conducted stated that the prevalence of psychosocial problem 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 somatization, grief, hopelessness, anxiety, inability to cope with anxiety, frustration, hostility, fear of death, lack of confidence, decreased self-esteem, loneliness, isolation, irritability, helplessness, lack of self-respect and feeling of lovelessness (15-17).

TRNC is a Mediterranean island that receives a large number of immigrants from other Mediterranean countries (18). Thalassemia is an important hematologic disease for the TRNC. Although no baby was born with Thalassemia on the island after 2001, the number of patient's present is substantially high. According to the data obtained from the Cyprus Thalassemia Association (2020), there are 87 patients with thalassemia major in TRNC. These individuals have many problems due to the reasons mentioned above. Since TRNC is an island that receives immigration, it has a sociological texture that includes different cultures, which can increase the dimension of this problem. For this reason, this review intends to examine the process and psychosocial aspects of major thalassemia, which is such an important disease for the TRNC.

Thalassemia in TRNC

The historical process of thalassemia in the TRNC includes prevention programs, education and awareness studies, screenings and genetic counseling and early diagnosis studies.

Thalassemia Prevention Program

It was first brought up in 1976 that thalassemia poses a serious public health problem in the TRNC. Studies toward the solution of this problem have been initiated throughout the country and the "Thalassemia Prevention Program" has been created and put into practice. Educational and social awareness studies have started since 1978 within the framework of this program. Risky families have been screened since 1979, and pre-marital screening tests and genetic counseling services have been provided to carriers since 1980. Prenatal early diagnosis studies were started in 1984 with fetal blood sampling, and chorionic villus biopsy and DNA studies have been continued since 1991. The conventional treatment methods recommended in the treatment of patients with thalassemia were applied and the life expectancy and quality of the patients were tried to be improved (19).

TRNC started first studies regarding thalassemia in different departments of Dr. Burhan Nalbantoğlu State Hospital, and gathered all services under the same roof with the establishment of Thalassemia Center in 1988. Thalassemia center consisted of clinical departments where daily follow-up and treatment were performed on patients with thalassemia, laboratories where screenings and early diagnosis studies were performed, and a blood bank. The Thalassemia Society office is also located in this center. The day care and treatment services are carried out by a hematologist, an internal medicine specialist and nurses trained in transfusion in the thalassemia center. In addition, treatments are planned and applied in cooperation with specialist physicians for the complications that develop in patients with thalassemia as the age increases (8).

Education and Social Awareness Studies

Public awareness and education are very important in fighting against this disease. In this framework, when the Thalassemia Prevention Program was initiated, effective training studies have been initiated in order to raise social awareness since 1978. Programs related to thalassemia were announced to the public through the press and broadcasting, conferences were held in schools, military units and universities, brochures and booklets on the subject were distributed to the public. Gynecology and pediatric specialists, general practitioners, midwives and nurses also participated in the training activities (19).

The most important country policy was to prevent births of sick children in the fight against thalassemia in the TRNC, especially by screening people of marriage and reproductive age and providing genetic counseling to those detected as carriers. In this context, before the start of the studies, circulars were sent to all obstetricians about the routine thalassemia screening of pregnant women who got married before 1980, so this group was also taken under control. Despite all these controls, it was thought that training and screening only risky groups would not be sufficient to prevent births of sick babies due to the high incidence of thalassemia on the island. For this purpose, pre-marital screenings were made legally mandatory in 1980 and it was decided that all couples to be married would be screened free of charge under state control. These practices were widely accepted by the public as well as supported by the State, and no baby with thalassemia was born after 2001 (8,19). In this context, considering that the prevention of a disease, the follow-up, and rehabilitation of the patients are very important in preventive health services, TRNC has taken an important problem under control successfully and continues its services.

Screenings and Genetic Counseling

The screenings were first started in Nicosia in 1978, and then continued in the Thalassemia Center for those who had a child with thalassemia in their family. Around 4500-5000 people were screened annually with the legal requirement to have a pre-marital thalassemia test since 1980, and these practices still continue in the country. Following the screening tests, genetic counseling is provided to couples diagnosed as carrier, and it is recommended to have an early prenatal diagnosis in case of a pregnancy. Some couples, who were both identified as carriers of thalassemia, refused to marry because they could not risk having a sick child in the first years of the program. People have become more conscious with the advancement of prenatal early diagnosis techniques, protection and awareness trainings, and such behaviors are no longer observed in the following years (19).

Prenatal Early Diagnosis Studies

One of the most important issues aimed was to prevent the birth of newborns with thalassemia within the framework of the program to fight against thalassemia in the TRNC. In this context, it was aimed to provide genetic counseling and prenatal early diagnosis services to all and to explain the risks of having a baby with thalassemia at a rate of 25% at each birth to couples who were both carriers. Prenatal early diagnosis studies have been conducted in line with these goals in the Thalassemia Center since 1984. During early diagnoses made by taking blood from the baby's umbilical cord with cordocentesis method, when the fetus was diagnosed with major thalassemia, these pregnancies were terminated with the consent of the family. With the introduction of DNA methods in 1991, earlier method was abandoned. Thus, early diagnosis studies were initiated and continued by examining the DNA of tissues obtained by chorionic villus biopsy at the 10-12th day of pregnancy (19).

Before the prenatal diagnosis studies, the number of new-born patients with thalassemia major was expected to be around 18-20 per year in TRNC. This number has decreased by approximately 50% with the initiation of prenatal diagnosis studies in 1984. Thus, the births of 600 new patients with thalassemia were prevented through screening, genetic counseling and prenatal early diagnosis studies in the last 30 years (20). These efforts to prevent thalassemia were first approached to raise awareness in the society, and no social resistance was encountered in the TRNC. The right health policy of the state has successfully reached the public, including the risky families. People approached these suggestions positively, and there were not many couples who did not accept early prenatal diagnosis. Around 10 babies with thalassemia were born for various reasons between 1984 and 2001, but no babies with thalassemia were born since 2001 (19). However, existing patients and carrier individuals who want to have children should be handled and followed very well in terms of psychosocial aspects.

Results of Thalassemia Treatments

It was aimed to fight against thalassemia, prevent the birth of sick children, and improve the life expectancy and quality of existing individuals with thalassemia by treating them in better conditions in our country. These studies started toward the end of the 1970s and continued intensely in the 1980s. Since it is a hereditary chronic disease, it is not possible to cure thalassemia by conventional methods. For this reason, treatments for the complete cure of the disease are performed with hematopoietic stem cell transplants in major centers around the world. It is essential to find stem cells compatible with one of the siblings in a large family for stem cell transplantation. Compatible stem cell supply between siblings is only successful at a rate of 30%. Studies have shown that conventional treatments have reached a quite good point on making the large patient masses survive longer and live better (21). In order to completely cure a patient's thalassemia in the TRNC, hematopoietic stem cells were taken from their sibling and transplanted to the patient, and the result was a successful. The patient who has reached adult age is currently living a healthy life (19).

Thalassemia treatment is carried out within the framework of the protocols below in TRNC (19).

• Adequate, effective and safe blood transfusions,

• Effective iron chelation, protection against infections, and regular vaccination,

• Follow-up and treatment for cardiological and hepatic complications,

• Tests and replacement therapies for growth and developmental retardation and other endocrinological complications,

• Tests and treatments for osteoporosis and bone metabolism,

• Screening against infectious diseases that can be transmitted by blood transfusion,

• Filtering the blood from leukocytes in order to prevent blood transfusion reactions,

• Detection of alloimmune antibodies that develop in patients against immunological transfusion reactions, and transfusion with appropriate blood,

• Psychological care and treatment for psychosocial problems.

Psychosocial Problems of Patients with Thalassemia Major

Patients with thalassemia major, experience physical and psychological problems. Patients experience many problems and mood disorders, including despair, sadness, hostility, depression, grief, anxiety, fear of death, hopelessness, social isolation, and helplessness (12,16).

Thalassemia disease also affects the hope of patients, as it is a disease that mostly affects the emotional state of the individual, changes the individual's perspective about the future and the world, affects life-related expectations, and autonomy. The scope of psychosocial care includes supporting and increasing hope and eliminating hopelessness in sick individuals. Hope is an important resource that helps people with thalassemia adapt to treatment and feel good. Health professionals should help the desperate individual to solve problems, develop their desires and make decisions easier, and instill hope in patients (9).

Professional healthcare team members have responsibilities such as education, care, counseling and organizing studies for individuals with chronic diseases. Individuals with thalassemia who are mental problems need professional help to cope with their problems. In such cases, it will increase the abilities of problem solving and decision-making, the development of wishes and desires of individuals, the development of positive patient-health professional relationship, the participation of the patient in treatment-related procedures, and the cooperation while solving problems (22).

There is an association of thalassemia patients in the TRNC. Patients and their families share their problems in social, psychological and therapeutic issues under the umbrella of the Northern Cyprus Thalassemia Association. As problems are shared, they become lighter and current shortcomings can be expressed in a stronger voice. The association also enlightens patients by raising awareness about the disease, following up-to-date treatments and innovations in this field. In addition, individuals with thalassemia take part in the management of the association personally and look after their problems (19). However, psychological support and treatment for psychosocial problems, which are also included in the thalassemia medical protocol, was given by a psychologist in the Thalassemia Center in the past years, but today there is no psychiatrist, psychologist, or psychiatric nurse in the treatment center in the TRNC. This situation is seen as a major deficiency by the patients (8). Therefore, a holistic approach should be adopted in the treatment protocol of patients with thalassemia, and the psychosocial problems of patients should not be ignored.

Conclusion

Services to redound the psychosocial care of patients with thalassemia are limited in the TRNC, and their treatment is carried out with blood transfusion and iron chelation. When the thalassemia process in the TRNC is examined, it is seen that thalassemia patients partly adapt to living with a chronic hematologic disease, but the psychological and social problems and treatment burdens they experience are at a level that should be taken into account. Therefore, healthcare professionals should pay attention to treatment and care for psychosocial problems as well as physical problems. Psychosocial care and support should be included in the treatment and care protocol of patients. In particular, nurses should support patients by providing psychosocial care.

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