T.C.
ÇANKIRI KARATEKİN ÜNİVERSİTESİ
SAĞLIK BİLİMLERİ ENSTİTÜSÜ
HEMŞİRELİK ANABİLİM DALI

QUALITY OF LIFE FOR PATIENTS WITH THALASSEMIA IN AL- NASIRIYAH CITY

Yüksek Lisans

Lara Abdulhsan Mathboob ALABOODI

Çankırı 2021

QUALITY OF LIFE FOR PATIENTS WITH THALASSEMIA IN AL-NASIRIYAH CITY

Lara Abdulhsan Mathboob ALABOODI

Sağlık Bilimleri Enstitüsü

Hemşirelik Anabilim Dalı

Yüksek Lisans

TEZ DANIŞMANI
Dr. Öğr. Üyesi Özlem BULANTEKİN DÜZALAN
İkinci Danışman: Dr. Alaa M. Tuama

Çankırı 2021

KABUL VE ONAY

Lara Abdulhsan Mathboob ALABOODI, graduate student of CHAKU, Institute of Health Sciences with the number of 198205312, prepared Lara Abdulhsan Mathboob ALABOODI,successfully presented his thesis titled “QUALITY OF LIFE FOR CLIENTS WITH THALASSEMIA IN AL- NASIRIYAH CITY” in front of the jury whose signatures are below.

Thesis Defense History:

Thesis advisor:	**Ozlem DUZALAN** **Cankiri Karatein University**	**……** **Signature**
Jury member:	**Name SURNAME** **……. University**	**……** **Signature**
Jury member:	**Name SURNAME** **……. University**	**……** **Signature**
Jury member: **(If applicable)**	**Name SURNAME** **……. University**	**……** **Signature**
Jury member: **(If applicable)**	**Name SURNAME** **……. University**	**……** **Signature**

I confirm the above result
Signature
Title Name SURNAME
Institute Director

ETHICS STATEMENT

The thesis titled “QUALITY OF LIFE FOR CLIENTS WITH THALASSEMIA IN AL- NASIRIYAH CITY” that I prepared and presented as a Master's thesis; Written by me in accordance with scientific morals and values. The idea/hypothesis of my thesis is entirely my own and my thesis advisor. The research in the thesis was done by me, and all sentences and comments are my own.

I declare that the above-mentioned points are correct.

Signature
27 July 2021
Lara Abdulhasn
Mathboob ALABOODI

DEDICATION

Her şeyden önce, merhametli ve şefkatli olan Allah'a çok şükür. Yrd. Doç. Dr. Özlem BULANTEKİN DÜZALAN. Danışmanım Danışman Dr. Alaa M. Tuama

CONTENTS

Page

ABBREVIATIONS AND SYMBOLS

HBF **:** Fetal Hemoglobin

PLT **:** Platlet

QOL **:** Quality of Life

RBC **:** Red Blood Cell

SCD **:** Sickl-Cell Disease

LIST OF TABLES

LIST OF FIGURES

AL- NASİRİYAH ŞEHRİNDE TALASEMİ'Lİ (AKDENIZ ANEMİSİ HASTALAR İÇİN YAŞAM KALİTESİ)

ÖZET

ALABOODI, Lara Abdulhsan Mathboob**.** Al-Nasiriyah Şehrinde Talasemili Hastalar İçin Yaşam Kalitesi, Yüksek Lisans, Çankırı, 2021

Talasemi, hemoglobini oluşturan alfa ve beta globin zincirlerinin normal üretimine zarar veren hemoglobinopati adı verilen bir grup genetik hastalığın bir parçasıdır. Latin Amerika ve Karayipler'de talasemi görülme sıklığı heterojendir. Sağlık çalışanları, hastalık ve tedavilerin etkileri hakkında bilgi sahibi olmaları nedeniyle, yetişkinliğe ulaşmış talasemi hastalarının bakımıyla yükümlüdürler. Talasemi majör için uygulanan yaşam boyu tedaviler, demir şelat tedavisi ile ilişkili düzenli kan transfüzyonlarını içerir. Palyatif olmasına rağmen, tekrarlanan kan transfüzyon tedavisinin uygulanması, hastaların yetişkinlik yaşına gelene kadar normal şekilde gelişmesini ve yaşam kalitelerini iyileştirmesini sağlar. Bu çalışma, Al Nasiriyah/Irak 2021'de talasemi hastalarına sağlanan yaşam kalitesi ve sosyal destek düzeyinin değerlendirilmesini belirlemek amacıyla yapılmış tanımlayıcı bir çalışmadır.

Deneysel olmayan bir tasarı olarak, örneklem 200 hasta ile olasılık dışı amaçlı veri türü kullanılarak gerçekleştirilmiş, hastalara sağlanan sosyal destek ve yaşam kalitesi konusunda anket kullanılmıştır. Veri tabanlarının ve istatistiksel tabloların geliştirilmesi için SPSS version 25 programı kullanılmıştır. Talasemi hastalarının yaşam kalitesi ile ilgili olarak, 200 hastanın %41.5'i yüksek yaşam kalitesine sahiptir Örneklemin %58'i sosyal desteğe bağlı olarak düşük yaşam kalitesi elde ederken, %46.15'i sosyal destek alırken, %53.84'ü sosyal destek almamaktadır. Hastanın yaşam tarzı ve hastalıkla mücadelesi değişmezse, kendisi için yüksek bir yaşam kalitesi elde etmeyecek ve bu da bu hastaların yaşam kalitelerinin düşmesine yol açacaktır.

Hastaların yaşam kalitesini yükseltmek için aile ve arkadaşlar tarafından sağlanan sosyal desteğin de artırılması gerekmektedir.

Anahtar Kelimeler: Talasemi (Akdeniz Anemisi) ile yaşam kalitesi, hastalar, transfüzyon terapisi

QUALTITY OF LIFE FOR PATIENTS WITH THALASSEMIA IN AL-NASIRIYA CITY

SUMMARY

ALABOODI, Lara Abdulhsan Mathboob. Quality of Life for Clients with Thalassemia in Al- Nassiriya City , (Master Thesis), Çankırı, 2021.

Thalassemia is part of a group of genetic diseases called hemoglobinopathy, which harms the normal production of the alpha and beta globin chains, which make up hemoglobin. In Latin America and the Caribbean, the incidence of thalassemia is heterogeneous. Health professionals face the care of thalassemia patients who have reached adulthood, due to advancing knowledge about the disease and the effects of treatments. Lifelong treatments for thalassemia major include the regular blood transfusions associated with iron chelate therapy. Although palliative, implementing repeated blood transfusion therapy enables patients to develop normally and improve their quality of life, until reaching the age of adulthood. this study determines the assessment of the quality of life and the level of the social support provided to thalassemia patients In AL Nasiriyah, Iraq 2021, and it is a descriptive study. As a non-experimental design, the sample was conducted using non-probability intentional data type with 200 patients, the questionnaire was used on the subject of social support provided to patients, and the quality of life. SPSS version 25 program was used to develop databases and statistical tables.

Regarding the quality of life of thalassemia patients, out of 200 patients (41.5%) obtained a high quality of life while (58%) of the sample obtained a low quality of life in relation to social support, (46.15%) received social support, while (53.84%) did not receive social support. If the patient's lifestyle and his coexistence with the disease do not change, a high quality of life will not be achieved for him, and this, in turn, will lead to a deterioration in the quality of life of these patients. The social support provided by family and friends must also be increased in order to improve patients' quality of life.

Key Words: Life quality, With thalassemia (Mediterranean anemia), patients, Transfusion-Therapy

1. INTRODUCTION

1.1.History of Thalassemia

Mediterranean anemia, or thalassemia, was described about a century ago. However, this is an ancient disease, as evidenced by archaeological discoveries and DNA remains (Rond, 2016). Their first descriptions were provided in 1925 in the United States by the pediatrician Dr. Thomas Cooley and the physicians Riti, Gribi, and Micheli in Italy. In children of Mediterranean origin, Cooley has observed severe anemia, large spleen and bone abnormalities. Experts in Italy described the same condition, but it is milder in youth and disease (Mettananda et al., 2019)

However, the disease takes its name from Whipple, who was a doctor on Rochester. The term "thalassemia" refers to a disease that mainly affects the population (Weatherall, 2004). The occurrence of the disease in the inhabitants of the Mediterranean Sea gave rise to detecting the causes and developing treatment methods. In 1942, an abnormal hemoglobin structure has been identified as the cause of Mediterranean anemia and anemia induction mechanism has been described. The body reacts by destroying them Red blood cells thus cause anemia. To make up for the deficiency , the body increases the speed of red blood cells, which causes the bones to challenge malformations, an enlarged spleen, and heart problems (Scherzadvir and Mukhtari, 2018).

In 1960, blood transfusion was proposed as a treatment for this disease. I found however, large amounts of iron were concentrated in the blood, which led to this the death. Thus, deferoxamine was discovered that prevents excess accumulation Iron in blood and prolongation of maximum life (Shirzadfar & Mokhtari, 2018).

1.1.1 Thalassemia in Iraq

The World Health Organization has indicated that about (35%) of non-pregnant women of childbearing age have anemia, and children under five years of age form (55.9%) and (38.2%) have anemia out of the total number of pregnant women. A proportion (25%) of breastfeeding women (IFHS 2006), and as noted from the above percentages, anemia is a severe health problem in Iraq, so it is important to reduce these rates to acceptable levels and to identify the most important health problems that lead to infection with this disease and deal with them in a Prompt and correct manner.

Also, this disease (anemia) must be addressed, its effects and its health and economic complications in Iraq, where anemia leads to slow growth, an increase in maternal mortality, a high percentage of premature children, and congenital deformities caused by a deficiency of folic acid. and physical businesses from 30 - 12%, as the United States of America alone loses 50 billion dollars annually from the gross domestic product.

In view of the importance and the economic and health effects of anemia, this study was conducted to identify its prevalence rates in the Iraqi society and for different age groups and in pregnant and lactating women and to investigate the most important factors directly related to the disease to direct health, human and financial efforts and resources to reduce the rates of this disease in the Iraqi society to the acceptable proportions approved by the World Health Organization.

1.2. Thalassemia (Mediterranean Anemia) Types

Mediterranean anemia is an inherited genetic disorder caused by the attenuated synthesis of one or more hemoglobin globin chains (Scherzadvir and Mukhtari, 2018). Hemoglobin consists of two pairs of chain polypeptides that are intertwined and give them a spherical shape. This structure of hemoglobin allows oxygen molecules in each chain to be bound, and then transported to the lungs and other tissues in the body. The type of hemoglobin is determined by the formation of the four chains. Fetal hemoglobin HbF contains two α chains and two chains, fetal hemoglobin α HbA contains two α and

two, with hemoglobin A2 having two α chains and two d. At birth, the proportion of hemoglobin HbF is 80% and HbA2 hemoglobin is 20%(Scherzadvir and Mukhtari, 2018).

1.2.1. A - Mediterranean anemia

It occurs as a result of a decrease or absence of synthesis of the alpha-globin chain. The production of α-globin is controlled by two genes, HbA1 and HbA2, on each chromosome 16. Simple gene deletion leads to the development of asymptomatic alpha-thalassemia while the test results are normal. However, if two genes are deleted, it causes alpha mesenchymal anemia Small cell anemia usually does not appear to a person. However, if there is a deletion of three genes, hemoglobin produces HbH, which contains four chains and is then stimulated, and is called smaller anemia, splenomegaly and hemolysis. If four genes are omitted, hemoglobin BART produces and causes fetal ascites which is fatal (Musallam et al., 2008).

1.2.2.B - Mediterranean anemia

Mediterranean anemia is caused by a deficiency or absence of-chain synthesis resulting in an increase in α-globin chains. Synthesis of β-globin controlled by one HBB gene on each chromosome 11, simple deletion of the gene has the same effect resulting in no symptoms or mild anemia. However, two gene deletions lead to a moderate to severe decrease in the production of the β-globin chain, leading to symptoms of moderate anemia. Deletion of three genes leads to the development of Mediterranean anemia, where symptoms appear from the sixth month of life, as in the previous period, the protective role of HbF. If reducing strings is not possible,

it causes moderate Mediterranean anemia, in which the individual's health status is taken into abetter ccount, as it does not require blood transfusion (Rund &Rachmilewitz, 2005).

1.3. Thalassemia Prevalence

Thalassemia is the most common recessive chromosomal disorder worldwide. The highest rate was recorded in the Mediterranean countries, in the Middle East and Southeast Asia (Mettananda et al., 2019).

The incidence of alpha + (mild) anemia characterized by the absence of two genes HBA1 and HBA2. , is high throughout tropical regions from Africa to the southeast while Asia α0 (severe) Mediterranean anemia, characterized by its complete absence of α-globin chains, is more common in Mediterranean populations and in southwestern Asia (Wirel, 2018).

Anemia in the Mediterranean region is more common in Southern Asia. Most of the reported cases of Mediterranean anemia are in Cyprus, Sardinia and Asia (Mettananda et al., 2019)

In contrast to these regions, where the disease prevalence is high and low, the incidence rate is in the United States of America and northern European countries. The incidents recorded in these areas come from immigrants from the above-mentioned countries (Weatherall, 2018).

1.4. Diagnosing Anemia

In the Mediterranean region, diagnosing anemia can become an easy task given that the fact that it can be performed through laboratory blood tests. An examination in which hemoglobin is separated by electrolyte can be given Answer. Determination of the mean hemoglobin volume is often the first indication of the presence of the disease, with a decrease in the mean size indicating its presence (Domars, Baum, Ekman, Giardina, Lynn & Schaeffer, 1996). If necessary, molecular DNA analysis is also performed (Rund, 2016).

Prenatal screening has played an important role in the diagnosis, as non

Invasive methods can be diagnosed. If there is a small amount of fetal material, blood is withdrawn from the mother and the diagnosis is possible (Rond, 2016).

Preimplantation genetic diagnosis can also be performed with the IVF procedure allowing diagnosis before the embryo implantation (Rond, 2016).

1.5. Thalassemia Treatment

The treatment for thalassemia depends largely on its severity. In previously reported cases of severe anemia, blood transfusions become imperative. However, blood transfusions have consequences for concentrating large amounts of iron in the blood. For this reason, drugs such as deferoxamine, deferiprone, and deferasirox are administered. Detail treatment medicines are a treatment for iron depletion, which greatly reduces the risk of premature death from cardiac toxicity due to iron overload in the heart. Desferrioxamine is given either by intravenous infusion or via a portable pump device. A dose of 30-60 mg / kg per day is injected into the body for 5-6 days. Limiting the active substance of 100 mg / kg can bind 8 mg of iron. However, the
The side effect from its administration is local irritation at the site where it occurred
infusion and fever. Long-term use causes ototoxicity Eye toxicity (Hashim & Mutlag, 2017).

Deferiprone is taken orally at a dose of 75 mg / kg in 3 divided doses, with each dose taken before meals. Although it is easy to use and increases patients' compliance with treatment it may cause arthropathy and agranulocytosis. Causing these side effects makes it necessary to stop treatment. Given the fact that deferiprone can lead to liver enzyme degradation and zinc deficiency, as well as serious consequences for the patient's health (Galanello & Campus, 2009).

Deferaxirose is taken orally and the recommended dose is 20-40
Mg / kg once daily, before meals, with chelation / cauterization for 24 hours (Cappellini, 2008). The side effect from its use is increasing the liver Enzymes and creatinine levels increase as well as digestive disorders (Cappellini et al. 2006).

It should be noted that the combination of deferoxamine and deferiprone can take place, it contributes to lowering iron levels in the blood while reducing its frequency with pump use (Origa et al., 2005).

In addition to blood transfusions and iron-deprivation therapy, overdoses can be used as treatments. One of these is the removal of the spleen, which can reduce the need for blood transfusions. However, the decision to perform a splenectomy is not an easy

matter, especially in patients with moderate anemia who do not have signs of splenectomy. The splenectomy procedure can be fatal, but the infections are considered before a splenectomy procedure (Graziano et al., 1981).

Despite the difficulty of the decision in many cases, it is considered necessary to understand, as the body of patients with moderate anemia is burdened with excess iron in the blood, with the risk of complications in the heart, in which several cases can be fatal. Splenectomy can not only reduce the need for blood transfusions, but also reduce the amount of deferoxamine the patient received. The complications of splenectomy can be reduced after surgery, pneumococcal vaccination, preventive penicillin and immediate treatment in case of infection (Graziano et al., 1981).

Another treatment is hematopoietic cell transplantation, where it is given to the non-allograft of the patient's allogeneic project, through which the blood disorder can be corrected.

To replace erythema is ineffective for the patient. For a transplant, it is necessary to find a donor compatible with the tissue. However, unrelated and unrelated volunteers are used to detect cord blood or incompatible relatives. After a successful transplant, patients do not need a blood transfusion. However, even if the first implant failed, he kept the second place. In general, it is recommended to perform this at an early age so that vital organs are not damaged. After transplantation, during the first 6-8 UTMs the bleeding should continue for months to remove excess iron from the blood (Ladis & Graphakos, 2011).

Finally, additional treatment can be genetic, which is a cause rather than a symptomatic treatment, as mentioned above. Attempt to focus on mutation in β-globin genes, DNA sequencing and RNA translation to repair-chain (Breda, Gambari & Rivella, 2009). They are the functional vectors of gene therapy Lentivirus vectors and these viral vectors are integrated into non-primary target cells of hematopoietic cells (Georgomanoli, Drakopouli, Papanikolaou & Anagnou, 2011). Now, the gene therapy available is the drug under the brand name Zynteglo, which consists of CD34 + cells, which contain hematopoietic stem cells, which are transformed by a lenticular vector that encodes the-globin gene (Scherzadvir and Mukhtari, 2018).

1.6 Thalassemia Prevention

Mediterranean anemia is a genetic disorder and prevention is the only realistic approach to birth control for people with Mediterranean anemia (Lucopoulos, 2011). But there is a huge role that pre-natal and preimplantation can play an important role in early diagnosis and genetic diagnosis. It is noteworthy that antenatal care can help couples who are likely to conceive a child with Mediterranean anemia to give birth to a child without Mediterranean anemia (Fesas, 1987).

1.7 Aims of Study

This study aim it :

- Assessing the transfusion-dependent quality of life for clients with thalassemia major
- Identifieying the relationship between the age of the patient, socioeconomic status of the family, social support and their quality of life dimensions, and patients' gender, marital status, educational qualifications and employment.

1.8 Problem of Study

Thalassemia has spread widely in the Mediterranean regions, especially in Iraq. The researcher chose the topic of thalassemia, the quality of life that a person lives with, and its relationship to some variables, due to the importance of the topic and the limited scientific research in this field in Iraq. The problem statement can be summarized with the following question:

What is the relationship of quality of life for thalassemia patients with the patient’s age, sex of the patient, the educational level of the father and mother, and the level of family income in the city of Nasiriyah, Iraq?

1.9 Limitations

- Time limits: 2021
- Place limits: the city of Nasiriyah in Iraq.

GENERAL INFORMATION

2.1 Alpha Thalassemia

IT is an inherited blood disorder that leads to impaired production of hemoglobin, the molecule that transports oxygen in the blood. Normal hemoglobin consists of two alpha chains and two beta chains; Quantitative reduction in alpha chains; Alpha thalassemia is the thalassemia in the HBA1 and HBA2 genes (Cheerva et al. , 2020). Alpha thalassemia. In addition, alpha thalassemia results in instability of beta globin molecules, which leads to increased red blood cell destruction. The degree of impairment depends on the clinical phenotype present in the number of genes that are affected (Vichinsky, 2013) as shown in Figure (2.1)

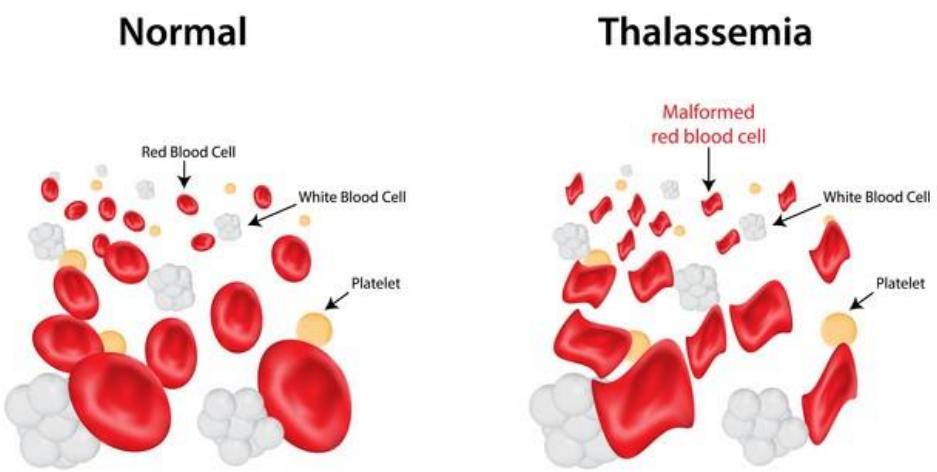

Figure 2.1: Normal blook and thalassemia blood (Pike and Bethesda, 2017)

2.1.2 Alpha Thalassemia Cause

Heredity, they also include deletions of 16p chromosome. in extreme circumstances , Alpha thalassemia may also be acquired (Tamary and Dgany, 2005)

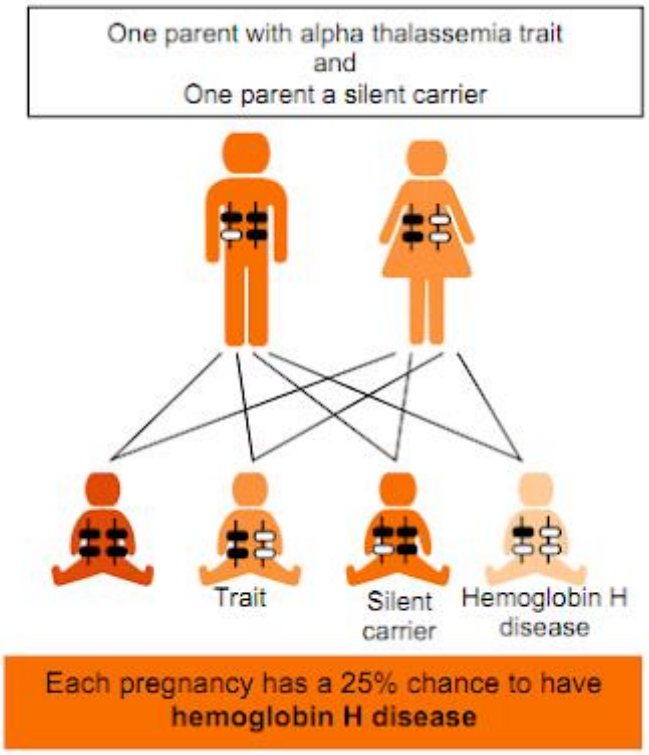

Figure 2.2: Thalassemia alpha by inheritance from father and mother (iAwesome,2012/ Available at : iawesome12.blogspot.com/2012/03/alpha-thalassemia.html)

The usual cause of alpha thalassemia is the deletion of the HBA1 and HBA2 genes. These two genes are used to make a protein called alpha-globin, which is part of hemoglobin (Robin and Miller, 2015).

All the human race has two copies of the HBA1 gene and each cell has two copies of the HBA2 gene. A single allele indicates which copy. One alloy is inherited from the father of one person for each chromosome, and the other from the human mother. As a result, four alpha globulins are synthesized. Loss of some or all of these diamonds results in different types of alpha thalassemia (Vichinsky ,2013).

The loss of all four alpha globin alles causes HB Bart syndrome, the most severe form of alfa thalassemia. The loss of three alpha globin alles results from the HbH disease. In both cases, a deficiency of alpha globin stops the cells from forming normal hemoglobin. Instead, the cells produce abnormal forms of hemoglobin called hemoglobin Barth (Hb Bart) or hemoglobin H (HbH). These abnormal hemoglobin

molecules cannot effectively carry oxygen to the tissues of the body. Replacing normal hemoglobin with normal hemoglobin leads to anemia and other serious health problems associated with alpha thalassemia(Vichinsky , 2013).

For α globalin, there are two genotypes thus in diploid cells, there are 4 alleles. Motherland has two alloys and root two is paternal. The severity of thalassemia depends on the amount of alpha-globin alleles affected: the more severe is the condition , a genotype is observed, "α" indicates a functional alpha chain, and indicates a pathological chain (Bajwa and Basit, 2020) as in Table (1.1)

Table 1.1 Alpha functional chain(Vichinsky, 2013; Palis, 2007; Harewood and Azevedo, 2020).

Alleles of which are affected	**Overview**	**genotypes**
1	Alpha thalassemia has a silent effect on the synthesis of hemoglobin called alpha thalassemia. Three α-globin genes are sufficient to enable normal hemoglobin development and no clinical symptoms. It is caused by a deletion mutation or non-deletion (Vichinsky, 2013)	-a/aa
2	This condition is called alpha thalassemia, where two alpha genes allow for the nearly normal production of red blood cells, but it indicates mild anemia. In many cases, it is confused with anemia that occurs due to iron deficiency and is treated without iron (Palis, 2007). **It comes in two types:** - Alpha-thal-1 (- - / α α), obtained through a	--/aa or –a/-a

	process of deleting the association states that are independent of each of the alpha genes on the same type of chromosome. The percentage is higher for people of Asian origins if they are compared to the rest of the world population - Alpha-thal-2 (- α / - α), obtained through transient deletion of alpha genes; When it gets chromosomes of different types and homogeneous. The high incidence and rate among Africans are very high if compared to the rest of the world population (Palis, 2007).	
3	This condition is known as hemoglobin H, as the two hemoglobin numbers are unstable in the blood. The hemoglobin-H and quadruple chains are present in the human body. This disorder is called hemoglobin H disease. Both of these unstable hemoglobin have a greater affinity for oxygen than normal hemoglobin. Peripheral hypertension, as well as an enlarged liver and spleen, can occur with microscopic anemia in target cells and Heinz bodies (precipitated HbH). In childhood and early adulthood, a disorder is noted, anemia and an enlarged liver and spleen are noted (Harewood and Azevedo, 2020).	--/-a
4	This type is called major alpha thalassemia; So that the fetuses are edematous, but have little hemoglobin, and the hemoglobin that is present is all quadruple chains. Thus, all four alleles are affected, and it is possible that the fetus will not survive the pregnancy without entering the uterus. Therefore, the majority of infants who have this type of alpha thalassemia major are stillborn, but it	--/--

	is possible that they will be treated while they are fetuses through blood transfusions for them while they are inside the womb during pregnancy until birth, and after birth, a bone marrow transplant or operations may be performed. Continuous transfusion of blood (Harewood and Azevedo, 2020)	

2.1.3 Diagnosis in laboratory

It is done through a complete blood count and through the red blood cell index for initial laboratory diagnostics. The peripheral blood supply must also be carefully checked. To diagnose alpha thalassemia, the hemoglobin test is crucial because it determines the types of hemoglobin and the percentages present. There are several methods of studying hemoglobin, which include hemoglobin electrophoresis, capillary electrophoresis, and high performance liquid chromatography (Mckenzie, 2011).

DNA molecular analysis (DNA analysis) may in particular be used for diagnostic diagnosis of alpha-thalassaemia (deletions and mutations in one or two genes of alpha-globin) for the identification of alpha-thalassaemia carriers (Daniel and Sabath, 2017).

2.1.4 A treat

Alpha thalassemia can also be treated by blood transfusions, which may reduce hemoglobin symptoms. The choice of beginning blood transfusions depends on the seriousness of the disease (Amin, 2019).

In the cases of overactive or swollen spleen anaemia or when therapies with blood transfusions are not available, splenectomy is a potential treatment choice to increase global hemoglobin levels. Splenectomy is however prevented if other options are available due to an increased risk of extreme thrombosis and infection (Zhou et al., 2014).

In addition, gallstones can be a surgical problem. Secondary fever complications must be monitored, and most people survive without treatment (Kongsnoth , 2007).

In addition, a treatment (and treatment), which is best done at an early age, for stem cell transplantation should be considered. Other options are still being developed, for example gene therapy (Zhou et al., 2014).

2.2 Beta Thalassemia (β thalassemia)

It's a group of blood diseases that are inherited. These are forms of thalassemia caused by poor or missing beta hemoglobin-synthesis which leads from severe anemia to people with no clinical signs. They lead to varying outcomes. It is estimated that the annual global incidence rate is 1 in 100,000. Beta thalassemia is due to a beta hemoglobin or HBB defect. The seriousness of the disease depends on the mutation's nature (Dreuzy et al., 2016)

Blockage of HBB over time decreases beta chain synthesis. The body's inability to create new beta chains results in decreased HbA development. Reductions of HbA in red blood cells are commonly available leading to anemia of microparticles. Small cell anemia develops gradually for adequate red blood cell function in combination with the deficiency of the HBB protein. Because of this factor, the patient may require a transfusion to compensate for the blockage in the beta chains. Blood transfusions repeatedly cause severe iron overload problems (Bajwa and Basit, 2020).

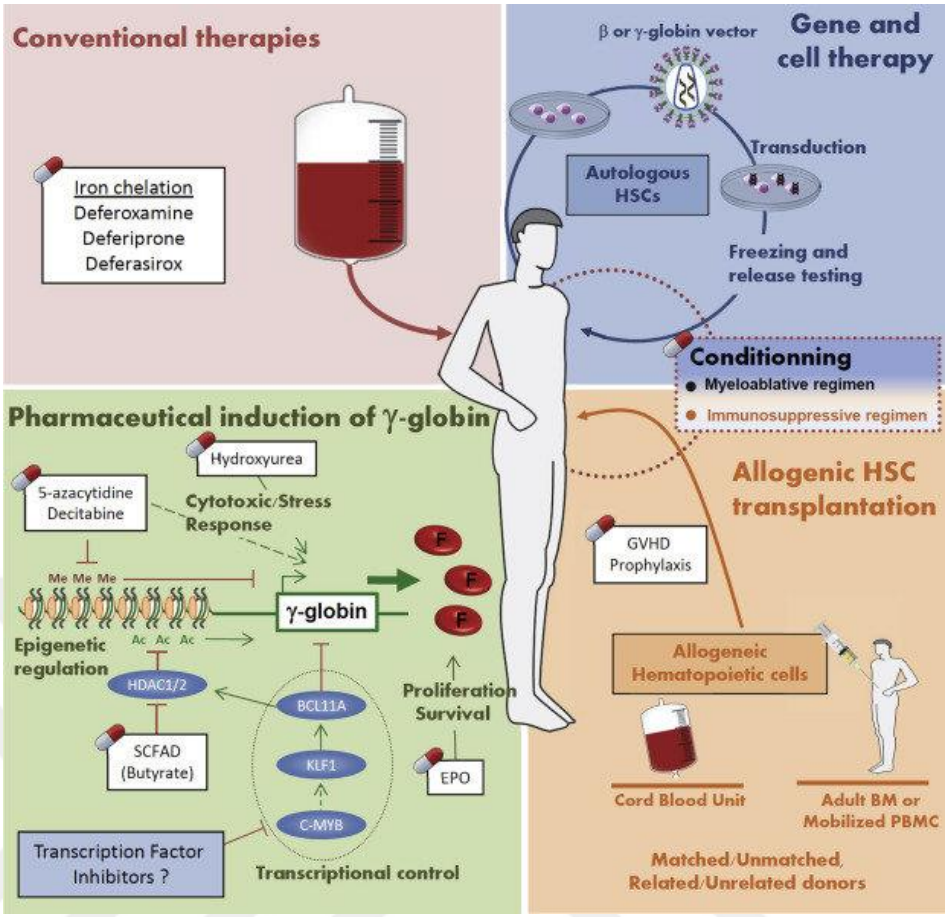

Figure 2.3: Beta thalassemia (Dreuzy et al, 2016)

Beta thalassemia has two main manifestations, which are classified according to the severity of symptomsinto : (Pike and Behesda, 2015)

2.2.1Thalassemia major

The more serious type, which, if left untreated, can cause severe anemia in an enlarged liver and spleen, is thalassemia major (also called anemia) (enlarged liver and spleen). By two years of age, this form usually becomes apparent. It causes failure to survive and low life expectancy if not treated. Treatment that is performed may be permanently canceled and sometimes cord blood may be used through a blood transplant or a bone marrow transplant (Herbert et al., 2020).

2.2.2 Thalassemia intermedia

The less serious type, which later becomes evident, causes milder anemia that does not require daily blood transfusions. Often, individuals with this type are at risk of iron overload (Herbert and Muncie, 2020).

Beta-thalassemia is caused and is usually inherited in an autosomal recessive manner by mutations in the HBB gene. This means that in each of their copies of the HBB gene, people with thalassemia major or thalassemia intermediate have a mutation (Benz, 2018).

2.2.3 β thalassemia causes

HBB gene mutations cause beta thalassemia. Instructions for producing a protein called beta-globin are given by the HBB gene. A part (subunit) of hemoglobin is beta-globin. There are four protein subunits of hemoglobin, usually two beta-globin subunits and two subunits of another protein called alpha-globin (Bethesda, 2020)

Some of the HBB gene mutations prevent any beta-globin from being created. HBB mutations in the gene cause beta thalassemia. The HBB gene provides instructions for making a protein called beta-globin. Beta-globin is a component (subunit) of hemoglobin. There are four hemoglobin protein subunits, typically two subunits of beta-globin and two subunits of another protein called alpha-globin (Thein, 2013).

Any of the HBB gene mutations prevent the development of any beta-globin. Absence of beta-globin is referred to as thalassemia beta-zero (β0). Other mutations of the HBB gene allow the development of certain beta-globin but in reduced quantities. A reduced volume of beta-globin is called thalassemia beta-plus (β+). However, possessing either β0 or β+ thalassemia does not necessarily predict the severity of the illness. However, having either β0 or β+ thalassemia does not generally predict the severity of the illness; individuals of both forms have been diagnosed with major thalassemia and moderate thalassemia (Bethesda, 2020)

There is a decreased amount of usable hemoglobin due to a lack of beta-globin. Red blood cells don't grow normally without adequate hemoglobin, creating a shortage of mature red blood cells. In people with beta thalassemia, the reduced number of

mature red blood cells contributes to anemia and other related health issues (Bethesda, 2020) as in figure (3.3)

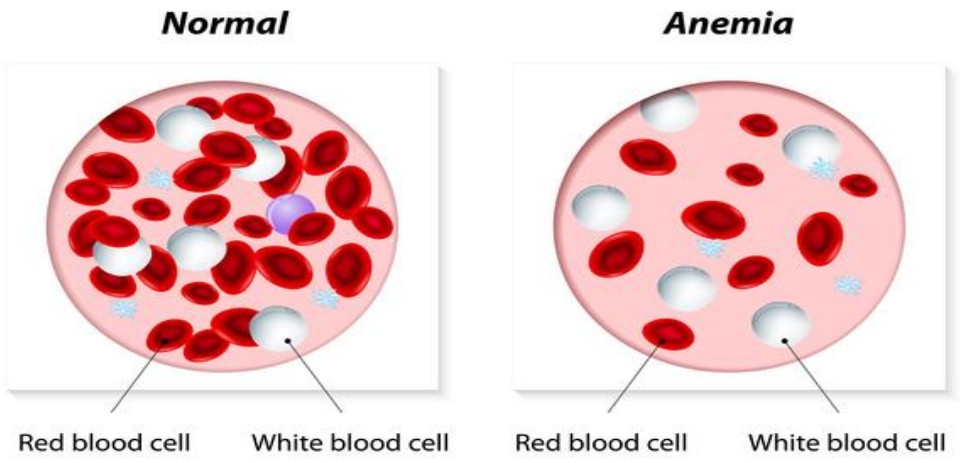

Figure 2.4 Normal blood and B Thalassemia blood (Bethesda, 2020)

2.2.4 Infection factors

Factors that raise the risk of developing beta thalassemia are family history and ancestry. Depending on family history, whether the parents or grandparents of a child have significant or intermediate beta thalassemia, there is a 75% (3 out of 4) risk of an offspring inheriting the mutated gene (Pinto and Forni, 2020). Even if a child does not have severe or moderate beta thalassemia, he or she could still be pregnant, which may lead to beta thalassemia occurring in future generations of their offspring. Proportions are another risk factor. In people of Italian, Greek, Middle Eastern, South Asian and African descent, beta thalassemia occurs most frequently (Pinto and Forni, 2020).

2.3 Organic Effects of Disease

2.3.1 Organic consequences of anemia in the Mediterranean:

Inefficiency of erythrocytes in the bone marrow in association with increased peripheral hemolysis leading to anemia which is associated with accumulation of that iron causes complications in various body systems.

2.3.2 Endocrine disorders in Mediterranean anemia

Endocrine disorders occur in patients with moderate anemia, caused by an endocrine disorder. Their occurrence increases with age and more than one endocrine disorder can occur in the same person. The most characteristic feature is residual growth, hypogonadism, diabetes mellitus, hypothyroidism and osteopathy (Diller, Mystery, Burbridge, Bancham, Wright, D., 2014).

Residual growth in people with moderate anemia, there is a delay in physical development, especially at the age of 5-9 years which leads to short stature. This developmental disorder is attributed to both blood transfusions and iron deprivation treatments, as well as to iron overload in the liver and pituitary gland, but also to folate deficiency of vitamin D18 and Zn19. It is worth noting that growth hormone disruption does not occur in all patients with moderate anemia and short stature (Pharmaki, 2011).

Hypogonadism hypogonadism is characterized by the absence or discontinuation of dangers attributed to a defect in the pituitary gland, during puberty due to its large size, iron concentration and oxidative damage caused by free toxic iron. It is observed in 40--60% of patients with moderate anemia, although they receive iron-deprivation treatment and undergo blood transfusions. Insufficient secretion of LH, FSH affects the development of the testicles and uterus. This results in boys' testicles growing smaller than normal and appearing.

Impaired azoospermia and decreased libido, while ovarian growth in girls is impaired and menopause (Farmaki, 2011).

2.3.3 Diabetes

Diabetes are one of the major endocrine diseases that occurs in 20-30% of patients. Patients with moderate anemia as a result of damage caused by iron concentrations in the cells of the pancreas. Before the onset of diabetes, there is a period of high insulin resistance and hyperinsulinemia. Chronic insulin secretion can lead to beta cell deficiency (Li et al., 2014).

2.3.4 Hypothyroidism

Hypothyroidism is a condition in which the body is unable to produce enough thyroid hormone (T4) (Garber et al., 2012). The frequency of its occurrence varies from 5-30% depending on the treatment protocol followed on a case-by-case basis. In Mediterranean anemia, hypothyroidism can be caused by damage to the pituitary gland (TSH-secreting thyroid hormone-TRH) or hypothyroidism (American Society of Endocrinologists, 2012).

In people with moderate anemia, damage to the pituitary axis has been noted (Landau et al., 1993). Symptoms caused by it are cold intolerance, low body temperature, weight gain, mood disorders, and fatigue. Hypoparathyroidism and hypocalcemia occur in 2-10% of patients with acute hemodialysis due to hypocalcemia documented through laboratory tests. Symptoms of hypoparathyroidism: Latent tetany (uncontrolled muscle contractions accompanied by hallucinations) and the presence of signs chvostek, trousseau. It is worth noting that it is not treated with hormonal therapy (Pharmaki, 2011). Hypocalcemia is associated with the appearance of heart complications, such as heart failure, which cannot be controlled with medication. Monitoring the patient's progress can prevent hypercalcemia that can lead to intracranial calcifications and hypercalciuria that lead to kidney stones (Demosthenous et al., 2019).

Orthopedic treatment is a common complication in adult patients with moderate anemia, regardless of gender. Ineffective redness of the bone marrow is "the main cause of bone breakdown, while iron deposition in the bone is toxic to osteoblasts and prevents bone maturation, leading to local ossification." High-dose treatment of desferoxamine, zinc, and vitamin D deficiency associated with initiation

Osteoporosis. Therefore calcium and vitamin D supplements are recommended but monitor the patient. Fractures due to bone can be prevented by proper nutrition, smoking cessation and increased exercise empowerment (Pharmaki, 2011).

2.3.5 Cardiac complications

Heart failure: Increased cardiac output is particularly common in patients with Mediterranean anemia. It is caused by anemia, but also by the compensatory

mechanisms used. It is characterized by dilation of the heart cavities, eccentricity of the left ventricle, normal or increased contraction, and diastolic filling corresponding to tumor overload.

In the past, even before a blood transfusion was treated, many patients with moderate anemia had died in the second decade of their lives due to heart failure (Pharmakis, Tirones & Aesop, 2011).

2.3.6 Cardiomyopathy

Cardiomyopathy results from an excessive concentration of iron and is characterized by the diastolic function of the left ventricle, while then it develops with “dilation and reduction of contractility of the left ventricle”, resulting in the appearance of dilatation

Restrictive cardiomyopathy occurs in a small number of patients "without dilatation or reduction of left ventricular contractility but of the restricted-diastolic type" (Farmakis, Tsironis & Aesop, 2011).

In addition to left ventricular stress, excess iron can lead to right ventricular cardiomyopathy and thus in manifestations of peripheral congestion (Cremastinos et al., 2010). Pulmonary hypertension Heart failure may occur in patients with moderate anemia, leading to increased pressure in the left ventricle of the heart and transmitted to the pulmonary veins and arteries. At the same time, symptoms of high blood pressure, functional and structural damage to blood vessels, hypercoagulation as well as respiratory infections can appear. The clinical manifestations of pulmonary hypertension are fatigue and shortness of breath, but also the cause of peripheral congestion due to curvature of the right abdomen (Du, Roguin, Milgram, Saab, & Koren, 1997). Arrhythmias Arrhythmias occur due to high iron concentrations. However, in the case of young patients, age must be taken into account

The likelihood of alcohol or other substance abuse, beyond thyroid hormones and its individual history (Taher and Capellini, 2018). 2.4 Hyperplenicity Inefficiency of erythrocytes leads to an overactive spleen. Anemia worsens and spleen hydremia develops (blood thinning) (Lucopoulos, 2015).

2.3.7 Bone abnormalities in persons with moderate anemia

Severe abnormalities of the Mongoloid facial bones and osteoporosis are observed. The fact that these are exhibited is usually attributed to the hypertrophy of the blood marrow caused by the absence of erythropoiesis (Loukopoulos, 2015).

2.4. QLO

It is a person's view of life, beliefs, aspirations, standards, and interests in the cultural sense as the quality of life (QLO) (WHO, 2006). However, thalassemia is well known, the positive effect of these treatments is likely to be decreased, particularly with regard to QOL, if it interferes with daily operations or is less likely to have an acquired blood condition that could be fatal when appropriate treatment does not occur. The life expectancy and survival of thalassemia patients have increased dramatically over the past decade through the introduction of regular transfusion therapies and iron chelate therapies. While patient survival improved

2.4.1. Health-related quality of life

In recent decades, the concept of quality of life has increasingly become an area of special interest for research in various fields. In the field of research on the quality of life is largely from a quantitative and qualitative point of view. Its importance lies in the fact that it provides a unified person-centered (or family-centered) structure, and a set of principles for increasing the subjective and psychological well-being of people (Borgna, 2004).

In the Handbook of Professionals in Education, Health and Social Services, the basic dimensions of quality of life are defined as "the set of factors that shape personal well-being". This guide identifies eight basic dimensions: emotional well-being, personal relationships, material well-being, personal development, physical well-being, self-determination, social inclusion and rights (Verdugo et al., 2005).

In the article the definition of QoL appears, which understands it as “the subjective evaluation that a patient makes of different aspects of his life in relation to his state of health”. These aspects are the physical, psychological and social aspects, disease symptoms and secondary effects of treatments (Eiser and Morse , 2001).

In 1994, the World Health Organization defined autobiography as "an individual's perception of his place in life within the context of the culture and the value system in which he lives and in relation to his goals, expectations, standards and interests."(WHO, 1994).

The World Health Organization's Quality of Life Toolkit (WHOQOL) also identifies a series of points, regarding health-related quality of life (HRQoL) measures, which have been accepted by different research groups. These measures should be: Subjective: Collect the perception of the person concerned. Multidimensional: It reveals different aspects of an individual's life, on the physical, emotional, social, interpersonal, etc levels (WHO, 2012).

- Including both positive and negative emotions.
- Recording the variance over time: age, stage of life passed (childhood, adolescence, adulthood, elderly), moment of disease occurrence, identify important differences in the aspects being evaluated.
- The WHO definition makes, in addition, a very valuable contribution, as it emphasizes the importance of cultural factors for self-evaluation (WHO, 2012).

In its Constitutional Charter of 1948, the World Health Organization proposed a broad definition of health as "a state of complete physical, mental and social well-being, and not merely the absence of disease." This definition has been questioned from different regions, some consider it impractical, others see it as a goal, and others consider it unattainable. However, it includes three characteristics that we can consider innovative: it reflects the concept of the individual as a person with a global focus, including a mental, social and environmental dimension, and links health with being productive and creative (WHO, 2018).

In 1977, the World Health Assembly set a goal in which all member states of the World Health Organization participate, and it is related to the progressive realization of a standard of health for all citizens of the world, allowing them to develop a socially and economically satisfying life. This strategy was defined in 1978 and confirmed by all member states of the World Assembly in 1979 (WHO, 2021).

It is also considered in the HRQL study as an evaluation of "the effects that disease induced and treatment have on patients' lives". This concept, although it is generally analyzed through a clinical post-treatment approach to evaluate the results of the interventions, it also allows for a very important analysis of not only the physical implications but also of the behavior and mood changes presented by the person with thalassemia (WHO, 2012).

2.5 The Effects of Mediterranean Anemia on the Individual and His Family

Thalassemia patients suffer from mental stress, as these tensions threaten and harm the mental and physical health of these people. Length of illness, duration of treatment, acceptance and increased cost of treatment, psychological state and social illnesses are those problems that affect a person and his / her family (Roy and Chatterjee, 2007).

Psychological problems have affected the quality of life (Origa, 2017). In the past, people believed that it was possible to provide an ideal condition through effective treatment and disease control. Today, however, evidence shows that we cannot achieve mental health and quality of life just by controlling symptoms. The pain experienced by a patient (such as thalassemia) is not only related to the vision of the nurses and doctors but also to the patient's personal feelings (Yu et al, 2019). Patients' responses to hospitalization vary, and some show negative reactions. The type and extent of these responses varies with regard to aspects of their personality, reaction time, etc. (Aljeesh, 2016). Therefore, many of these diseases need help in order to solve the patient's problems (Aljeesh, 2016).

2.5.1 Effects of Mediterranean Anemia on an Individual's Psychology

Mediterranean anemia is a chronic disease. It is persistent and treatment is used to manage the effects of Mediterranean anemia and prevent more serious complications (Di Matteo & Martin, 2002). The need for lifelong compliance with medical treatment in addition to frequent patient visits to the hospital to achieve the required transfers, as well as treatment to remove iron may affect their emotional state (Mednick et al., 2010).

The psychological effects of Mediterranean anemia began to appear since childhood, with anxiety and depression being the most characteristic (Messina et al., 2008). Children with beta thalassemia have a greater degree of mental disorders than those of children with other chronic diseases (Tsiantis et al., 1996). Preschool children with anemia in the Mediterranean tend to be more dependent on their parents compared to their healthy peers (Tsiantis et al., 1996).

The emotional difficulties they face due to the way they view themselves compared to them. Their peers differ from themselves in appearance while not needing to. They often miss school, just as they are. At the same time, they are encouraged to face the side effects of the recommended iron therapy, which cause fatigue and sometimes hinder their participation in various activities (Burhani, Najafi, Rapuri and Sabzivari, 2011).

Along with the difficulties that a chronic disease brings in their lives, family and social difficulties a child faces, but also genetic factors can lead to the onset of depression (Koutelekos & Haliasos, 2013).

It should be noted that in addition to depression in children in adolescence, provocative antisocial disorder is more common in boys (37%) versus girls (7%) (Beratis, 1993).

2.5.2 It’s main clinical manifestations

Tantrums are tantrums, arguments, and unnecessary harassment of others and holding others accountable for personal errors (Wenar & Kerig, 2000).

Relationships play a crucial role in the emotional state of their peers. It is well established that children are influenced by peers who may not accept or reject their

classmates, and who have a chronic illness. Especially children with moderate anemia are subjected to social stigmatization due to their disease (Georganda, 1988). However, the degree to which a child is chronically ill suffers rejection from his peers regarding his body status and whether he can participate in activities, such as his peers. Perhaps this is a factor that explains why their poor performance is also characteristic of chronically ill children who tend to be more shy due to the embarrassment caused by their illness and their treatment (Di Matteo &Martin, 2002).

2.5.3 Problems related to adolescence occur during adolescence

Delayed entry into adolescence. Adolescents with moderate anemia due to delay leading to short stature, delay in sexual development, as well as bone deformities, may experience discomfort.

Low self-esteem: At the same time, their emotional state is also affected by excessive parental protection. However, they try to become independent and take charge of their illness, which is passed on from parents to themselves (Angastiniotis, 2002).

But as adolescents get older, their levels tend to decline in depression, which can be attributed to the way they began to perceive their state of health and to form a mature perception about the impossibility of changing all situations that occur in life (Mikelli & Tsiantis, 2004).

As adults, they encounter difficulties related to their working life and finding a mate. Occupation selection is very important so that they can keep up with the requirements of each job, but also with the needs of the disease. They need to spend many hours on their treatment and often have to walk away from their work to attend their scheduled blood transfusion appointments. At the same time, they must disclose to employers their state of health in order to justify frequent absenteeism from work (Muslim, Capellini, and Taher, 2008).

To judge the psychological and social burden of patients with moderate anemia, it is necessary to provide psychological support. Its goals should be different for every age and clearly tailored to the needs of the patients. More specifically, during childhood, psychologists, as well as staff in units dealing with Mediterranean anemia, should aim

to understand the nature of the child's illness and try to manage his denial, so accept his health condition and be able to talk about matters that interest him (De Matteo & Martin, 2002) .

Intervention in adolescence should be carefully planned and targeted in adolescent participation in its treatment. His training in methods of managing problems that arise or will arise in the future are necessary in order to be

Psychologically prepared and empowered: The role of the psychologist is catalyst, as they can be taught the ways in which they will deal with their psychological difficulties, but also to help them have better academic performance (management exhaustion and lack of energy) and lead to suitable for themselves career choices (Muslim, Cappellini, and Taher, 2008).

2.5.4 Impact of Mediterranean Anemia on the Family.

In the past, the average life expectancy of patients with moderate anemia was low, it created anxiety and fear of death, not only for those afflicted but also for their families. Parents of these children, in addition to anxiety and fear of death, feel guilty about the state of their child's health. The severity of Mediterranean anemia in

Along with the requirements of adapting to the daily routine of frequent hospital visits, it burdens the family psychologically and financially (Sapountzi-Krepia et al., 2006). At the same time, it has an impact on their work lives and affects their personal relationships (Mastroyannopoulou, Stallard, Lewis, & Lenton, 1997).

As a result of this chronic disease and what it entails, they become overprotective parents of the child with moderate anemia and bear the responsibility of caring for him and complying with the recommended treatment. If there are other children in the family, it is very likely siblings are afraid of losing their sick brother as well as the parents, who have to go to hospital often with the child. However, the dynamics play a critical role in how the rest of the family members will manage the chronic illness of the patient (Di Matteo & Martin, 2002).

Families with strong foundations and their members have strong bonds between; they can better manage the situation. Conversely, in families with conflict resolution difficulties and who find it difficult to talk about a child's illness, the members become more psychologically burdened and alienated from each other (Shapiro, 1986).

3. METHODOLGY

This chapter deals with the methodology adopted for the study. It includes the research approach, research design, variables, setting, population, sample, and criteria for selection of the sample, sample size, sampling technique, development and description of the tool, content validity, pilot study, and reliability of the tool, data collection procedure and plan for data analysis.

3.1. Research Approach:

A quantitative research approach has been used for this study.

3.2 Study Design:

The study was a cross-sectional study conducted among adolescents (ages 12-20 years) with Thalassemia who received treatment at the Center for Genetic Blood Diseases, Nasiriyah city, in Iraq.

Describing the burden of disease in the community and its distribution. it can be a frequent measure of change in the population by collecting data mostly via questionnaire or by structured interview about more than one case at a time with the possibility of detecting correlation patterns between the pooled variables.

Moreover, it allows collecting large amounts of data from a large number of people in a variety of topics, so the data can be used by different researchers Majors. The cross sectional research also works well for exploratory studies.

3.3.Administrative Arrangement

Prior to the initiation of this study, approvals were obtained from the Behavioral Research and Ethics Boards of Cankiri Karatekin University to ensure on 20/10/2020

that this research study complies with the ethical standards and safety of the participants. After approval by the Behavioral Research and Ethics Councils, the researcher provided a detailed description that includes the aim of the study and its methodology to the Iraqi Ministry of Health to obtain official permission, and it was presented to Thi-Qar Health Directorate of Nasiriya . Sector, to ensure its approval and cooperation.

3.4 Choosing a Sample:

3.4.1. The inclusion criteria:

1. Thalassemia patients.
2. Patients (12-20 years).

3.4.2. Exclusion criteria:

1. Patients with severe disease that may limit their ability to participate in a study.
2. Patients who do not wish to participate in the study

3.5. Location and Population:

The participants were recruited from the Genetic Blood Diseases Center, Nasiriyah, Iraq.

3.5.1. Pre recruitment and recruitment:

Thalassemia patients were recruited while attending the clinic for routine follow up.in addition we used a social network to arrange appointments with thalassemia patients to discuss about our research project.

3.5.2. Sample size

We were intending to recruit all thalassemia patients who attended their clinic during the study, but only 200 participants were possible to be recruited in the study period.

3. 6 Data collection:

3.6.1. The procedure

The data were collected through the use of a questionnaire and through the method of interviewing the patients, then all eligible patients between (12-17 years) ages, their parents, and patients between (18-20 years) were contacted, as they came for routine follow-up during a period. Data were collected, and the Genetic Hematology Center, Nasiriyah, Iraq (study sample). The interviews were conducted in Arabic, which is the national language of Iraq. All data collected from the interview were documented using the hard copy method. The data collection process was conducted from September 2, 2020 until April 30, each patient spends between (25-30) minutes responding to the interview.

3.6.2 The Study Instrument:

A questionnaire was designed and constructed by the researcher to measure the variable. Such a construction was employed through the review of literature and related studies. The questionnaire consisted of 3 parts (Appendix C).

3.6.2.1 Demographic Data Form:

A demographic data sheet, consisted of (8) items, which included Age, Gender, Marital status, Educational qualification, Family's socioeconomic status, Level of Education for Father and Mother , Household's occupation and Family's Monthly Income (Iraqi Dinar).

3.6.2.2 Social Support Model

The second part of the questionnaire included (14) items related to Duke-UNC social support, and a list of some of the things that others do for us or provide for us that may be helpful or supportive. Such as visits with friends and relatives, help around the house, help with money and more

The third part of the questionnaire included (31) items related to transfusion-dependent quality of life.

3.7 Classification and Registration:

Items were classified and scored according to the following patterns:

- A five-point Likert scale: is used to classify items as never, rarely, occasionally, often, and always
- The five-point Likert scale is used as: 1 for Never, 2 for Rarely, 3 for Sometimes, 4 for Most, and 5 for Always
- Dependent variables:
- There are 8 different components of the questionnaire
- Independent variables:

Are categorical variables. Nominations of thalassemia patients according to the quality of life scores such as age group, gender influence, and education level.

3.8 Data Analysis

Questionnaires were completed and collected from the participants, and data was presented in the Excel file, SPSS software, using Windows version 26 and the Statistical Package for Social Sciences (SPSS), was used to analyze and compare the results of the current study.

Mean differences in the different components of the questionnaire with a previous study were conducted in the same center with identical specifications. Descriptive statistical measurement of frequency, percentage, mean, standard deviation, Pearson correlation, paired T-test, and analysis of variance (ANOVA) were performed.

3.9 Data Processing:

Questionnaires were collected and analyzed. The data have been saved in a hard copy. The confidentiality of the patients' details was preserved and the researcher was

only the one who could reach them in addition to the supervisors from the city of Baghdad who participated in the supervision of this research.

3.10 Conducting a Pilot Study:

A pilot study was performed on 10% of the sample (20 patients) to judge the feasibility of the study, objectivity, and testing of the tool's capacity. The sample was chosen by the Genetic Blood Diseases Center, Nasiriyah, Iraq, before the original study. In addition, Table (1-3) shows the determination of the sample that was selected according to its preparation. People who participated in the pilot study were excluded from the actual study. The pilot study was conducted from April 30, 2020 through May 15, 2020.

3.10.1 The purposes of the pilot study are:

- Determining the credibility of the researcher dependency (Inter Examiner) and determining the credibility of the patients (Intra Examiner) by testing retest.
- Confirming the clarity and adequacy of the content of the tool structure throughout the understanding of the topics and identifying the required modifications.
- Estimating the average time required to collect data for each patient during the interview process.
- Determining the best approach needed to know the nature of the difficulties they may encounter.

Table 3.1: Reliability Coefficients of the Pilot Study intra examiner and inter examiners

N	Setting	No.	%
1	Genetic Blood Diseases Center, Nasiriyah, Iraq.	20	100%
Total		20	%100

Table 3.2: Reliability Statistics using spss

Reliability Statistics				
Cronbach's Alpha		No. of Items		
.981		5		
	Scale Mean if Item Deleted	**Scale Variance if Item Deleted**	**Corrected Item-Total Correlation**	**Cronbach's Alpha if Item Deleted**
I had enough energy for daily activities	13.1800	28.249	.934	.980
I found it hard to keep up physical activity with my peers	13.4500	25.595	.965	.973
My health allows me to participate in as many social events as I would like	13.1700	24.805	.973	.973
Pain prevented me from doing what I need to do	13.7600	27.661	.896	.983
I felt too tired to do the things I enjoy doing (i.e., hobbies, crafts, sports, musical instruments)	13.4000	25.307	.972	.972

Table 3.3: The Duke-UNC Functional Social Support Questionnaire

	Relevant (Rating ≥ 3)	Irrelevant (Rating ≤ 2)	I-CVI	Interpretation	S-CVI
Item 1	11	1	91.7	Appropriate	83.9
Item 2	11	1	91.7	Appropriate	
Item 3	11	1	91.7	Appropriate	
Item 4	9	3	75	Appropriate	
Item 11	9	3	75	Appropriate	
Item 6	11	1	91.7	Appropriate	
Item 7	9	3	75	Appropriate	

Item 8	10	2	83.3	Appropriate	
Item 11	10	2	83.3	Appropriate	
Item 10	9	3	75	Appropriate	
Item 11	11	1	91.7	Appropriate	
Item 12	11	1	91.7	Appropriate	
Item 13	8	4	66.7	Appropriate	
Item 14	11	1	91.7	Appropriate	

Table 3.4: Statistics of reliability and credibility by 6 referees at University of Baghdad

	Relevant (ratings ≥ 3)	Irrelevant (ratings ≤ 2)	I-CVI	Interpretation	S-CVI
Item 1	12	0	100	Appropriate	99.5
Item 2	12	0	100	Appropriate	
Item 3	12	0	100	Appropriate	
Item 4	12	0	100	Appropriate	
Item 11	12	0	100	Appropriate	
Item 6	12	0	100	Appropriate	
Item 7	11	1	91.7	Appropriate	
Item 8	11	1	91.7	Appropriate	
Item 11	12	0	100	Appropriate	
Item 10	12	0	100	Appropriate	
Item 11	12	0	100	Appropriate	
Item 12	12	0	100	Appropriate	
Item 13	12	0	100	Appropriate	
Item 14	12	0	100	Appropriate	
Item 15	12	0	100	Appropriate	
Item 16	12	0	100	Appropriate	

Item 17	12	0	100	Appropriate	
Item 18	12	0	100	Appropriate	
Item 19	12	0	100	Appropriate	
Item 20	12	0	100	Appropriate	
Item 21	12	0	100	Appropriate	
Item 22	12	0	100	Appropriate	
Item 23	12	0	100	Appropriate	
Item 24	12	0	100	Appropriate	
Item 25	12	0	100	Appropriate	
Item 26	12	0	100	Appropriate	
Item 27	12	0	100	Appropriate	
Item 28	12	0	100	Appropriate	
Item 29	12	0	100	Appropriate	
Item 30	12	0	100	Appropriate	
Item 31	12	0	100	Appropriate	

Table 3.3: Table 3.4:

This is a testing of the study instrument validity, by 6 referees at University of Baghdad

4. Results and Findings

In this chapter, the results of the data analyzes will be presented. First, the socio-economic and demographic characteristics of the sample, followed by the social support for thalassemia patients in the study sample, then the quality of life scores for the patients to answer the research objectives and questions, the sample was selected at the Genetic Hematology Center in Dhi Qar Governorate, Nasiriyah Region, Iraq, using the method of random sampling. Simple (4.1) The demographic, economic and social characteristics of the sample: The total number of eligible patients was 200 patients with thalassemia Major only while two of the patients were diagnosed with thalassemia major and sickle cell anemia It is shown in Table 4.1

Table 4.1 : Distribution of the studied groups with comparisons significant (n: 200)

Basis Information	Groups	Frequency	Percent
Age Groups (years.)	12-14 years	57	28.5%
	15-17 years	62	31%
	18-20 years	81	40.5%

	Total	**200**	**100%**
Gender	Male	94	47%
	Female	106	53%
	Total	**179**	**100%**
Marital status	Single	200	100%
	Married	0	0%
	Total	**200**	**100.0**
Education Level	Sixth Primary School	84	42%
	First Middle school	11	5.5%
	Second Middle school	19	9.5%
	Third Middle school	22	11%
	Fourth Preparatory school	26	13%
	Fifth Preparatory school	9	4.5%
	Sixth Preparatory school	13	6.5%
	College First Stage school	8	4%
	College Second Stage school	8	4%
	Total	**200**	**100.0**

Table (4.1) The study indicated that the majority of the study sample (63%) are females and the rest are males, and most of them are between the ages of 20-18 years (40.5%). Only 57 patients were in the age group of 14-12 years (28.5%). Regarding the issue of marital status, all of the sample members were celibate and constitute (100%) of the total sample**.**

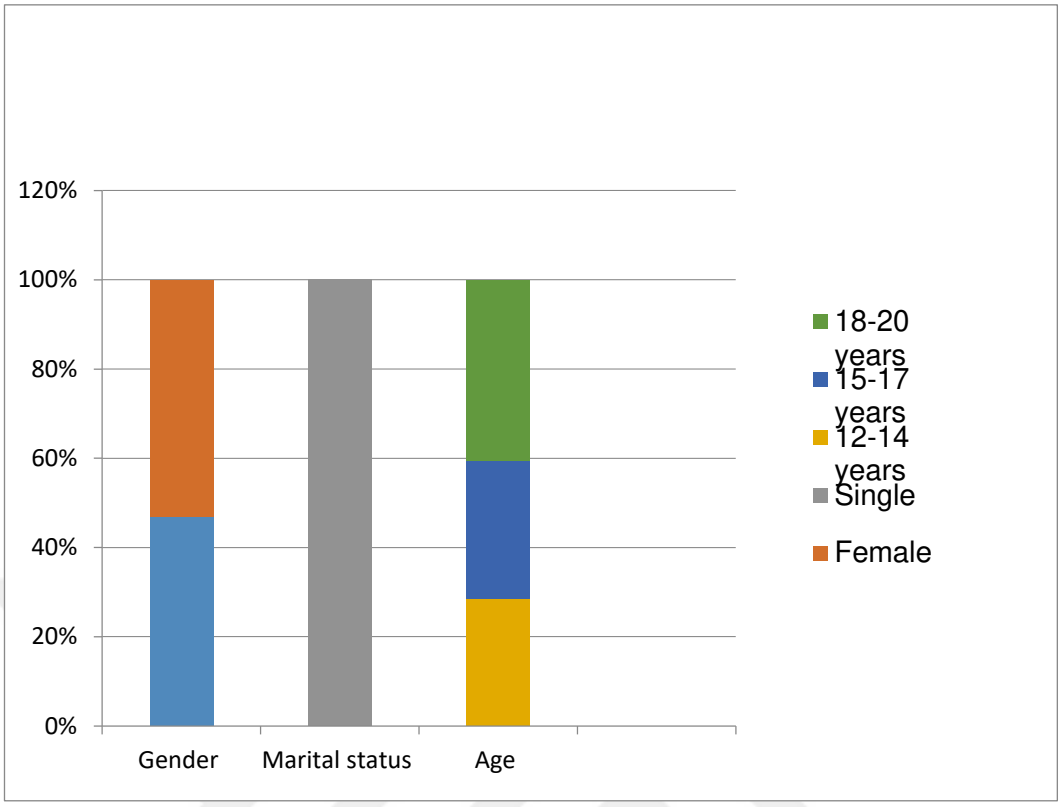

Figure 4.1: Demographic Characteristics Of Quality Of Life For Clients With Thalassemia in Al- Nasiriyah City, Iraq

Table 4.2 : Family's socioeconomic status for the studied groups with comparisons significant (n: 200)

Basis Information	Groups	Frequency	Percent
Householder's occupation	Professional	37	18.5%
	Semi-Professional	31	15.5%
	Clerical, shop owner farmer	29	14.5%
	Skilled worker	16	8%
	Semi- Skilled worker	14	7%
	Unskilled worker	73	36.5%
	Total	**200**	**100.0**
Family's Monthly Income Iraq diner	< 300.000	93	46.5%
	601.000-900.000	18	9%
	1.201.000-1.50.000	25	12.5%
	300.000-600.000	37	18.5%
	901.000-1.200.000	21	10.5%

	≥ 1.501.000	6	3%
	Total	**200**	**100.0**

Table (4.2) shows the socio-economic status of the family for the studied groups, as it appeared that the profession of the head of the household for the majority of the sample was at the level of the unskilled worker, as they formed (36.5%) of the sample members, while the work of the head of the household formed only (7%) at the level of the semi-skilled worker. The rate of (8%) was the skilled worker, as well as (18.5%) is at the level of the professional worker and (15.5%) is the level of the semi-professional worker, in addition to (14.5%) at the level of the clerk, the shopkeeper. As for the monthly income of the family, the largest number of them was (93) of the total sample, who received less than 300,000 Iraqi dinars, or (46.5%) from the sample members, meaning that they live with a low monthly income level, and 6 individuals from the sample received Only (3%) of them received an income of more than 1,501,000, i.e. with a high level of income, and a percentage (18.5%) of the sample obtained an income between 300,000 - 600,000 Iraqi dinars, and a percentage of (18.5%) also obtained (12.5%) on a monthly income estimated at 1,201,000-1.50,000 Iraqi dinars, i.e. with a somewhat average income level, and (10.5%) of the sample obtained a monthly income greater than 901,000 - 1,200,000 Iraqi Dinars

Table 4.3 : Family's education status for the studied groups with comparisons significant (n: 200)

Basis Information	Groups	Frequency	Percent
Father Education Level	Unable to read and write	16	8%
	Read and writhe	30	15%
	Elementary school	47	23.5%
	Middle school	39	19.5%
	High school	21	10.5%
	Diploma	19	9.5%
	Bachelor's	28	14%
	Total	**200**	**100.0**
Mother Education Level	Unable to read and write	34	17%
	Read and writhe	24	12%
	Elementary school	47	23.5%

	Middle school	27	13.5%
	High school	18	9%
	Diploma	22	11%
	Bachelor's	28	14%
	Total	**200**	**100.0**

As Table (4.**3)** for the educational level of the father, the largest number of them were from Elementary school graduates, as they constituted (23.5%) of the sample members, while 16 individuals in the sample did not read or write (8%) only, and (19) individuals received diploma education, i.e. a percentage. (9.5%) of the total sample, while the Middle School education received 39 members of the sample, i.e. (19.5%). Also, 15% of the participating parents were able to read and write, and secondary education received 21 sick parents i.e. By (10.5%), as for the educational level of the participating mothers, the largest number received education in the Elementary school by (23.5%) of the total sample of patients' mothers, and (9%) of the mothers received secondary education, while (34) mothers could not read And writing, i.e. (17%) of the sample, and (12%) of them could read and write. There were 18 mothers who obtained a diploma, or (11%) of the total number of mothers of thalassemia patients, and (28) mothers obtained a bachelor's degree.

Table 4.4 Distribution of the studied groups according to the results of social support with significant comparisons (n = 200)

Basis Information	**Groups**	**Frequency**	**Percent**	**Std.**	**Quality of life Level**
Visits with friends and relatives	As much as I would like	169	84.5	36.281	Supporter
	Much less than I would like	31	15.5		Not Supportive

Help around the house	As much as I would like	127	63.5	0.48264	Supporter
	Much less than I would like	73	36.5		Not Supportive
Help with money	As much as I would like	132	66	0.47490	Supporter
	Much less than I would like	68	34		Not Supportive
Praise for a good job	As much as I would like	182	91	0.28690	Supporter
	Much less than I would like	18	9		Not Supportive
People who Care	As much as I would like	157	78.5	0.41185	Supporter
	Much less than I would like	43	21.5		Not Supportive
Love and affection	As much as I would like	170	85		Supporter

	Much less than I would like	30	15	0.35797	Not Supportive
Telephone Calls	As much as I would like	171	85.5	0.35298	Supporter
	Much less than I would like	29	14.5		Not Supportive
Chances to talk-work	As much as I would like	124	62	0.48660	Supporter
	Much less than I would like	76	38		Not Supportive
Chances to talk-personal	As much as I would like	123	61.5	0.48782	Supporter
	Much less than I would like	77	38.5		Not Supportive
Chances to talk-money	As much as I would like	115	57.5	0.49558	Supporter
	Much less than I would like	85	42.5		Not Supportive

Invitation	As much as I would like	158	79	0.40833	Supporter
	Much less than I would like	42	21		Not Supportive
Useful Advice	As much as I would like	180	90	0.30075	Supporter
	Much less than I would like	20	10		Not Supportive
Help with transportation	As much as I would like	168	84	0.36753	Supporter
	Much less than I would like	32	16		Not Supportive
Help when Sick	As much as I would like	184	92	0.27197	Supporter
	Much less than I would like	61	8		Not Supportive

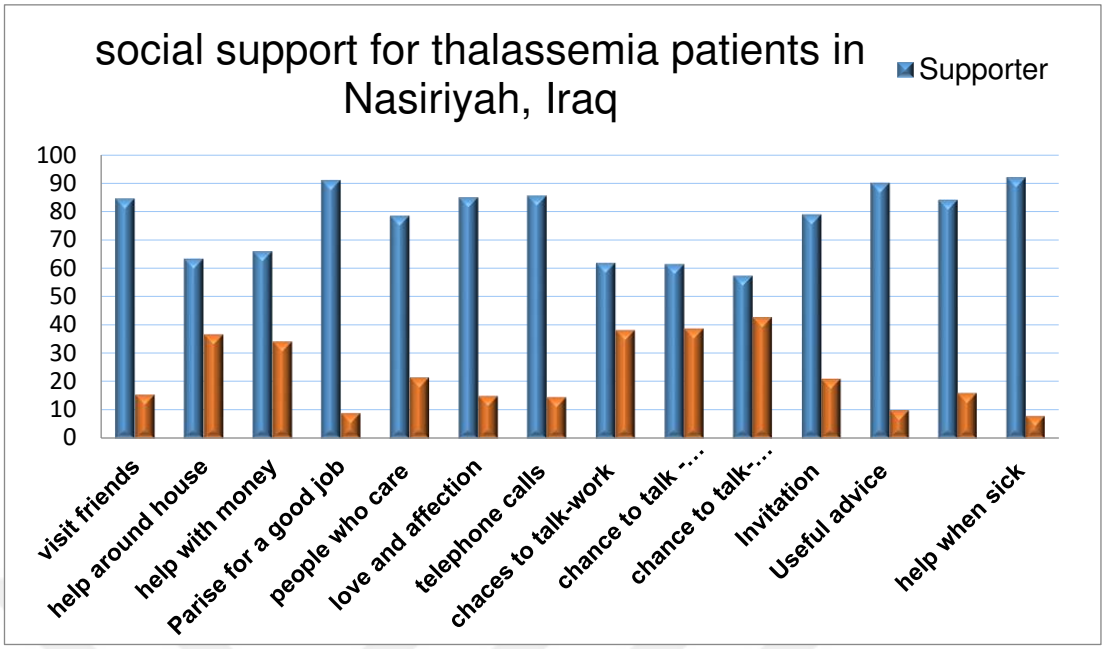

Figure 4.2: Social support for quality of life for thalassemia patients in Nasiriyah, Iraq

Through the results of Table (4.4) the results of social support for the studied sample of thalassemia patients were presented, we find that (84.5%) of the study sample received social support through visits with friends and relatives, and (15.5%) did not receive it. Help around the home: (63.5%) of the sample received social support and (36%) the social support was less than expected, and (66%) of the sample expressed social support in terms of financial aid, while (34%) was Unsupportive. He also received support through praise for a good job at a rate (91%), while only (9%) received it much less than expected, and social support was obtained through people's attention at a rate of (78.5%), but (21%). (5%) could not obtain it, and (85%) received social support through love and affection, and (15%) did not receive it and it was much less than expected. As for telephone calls, (85.5%) also got It is considered as social support, and the rate (15.5%) is much lower than expected.

A percentage (62%) received job opportunities and that was as social support, but a percentage (38%) received it much less than expected, and (61.5%) received social support through opportunities to talk about personal matters, while a percentage (61.5%) received social support through opportunities to talk about personal matters. (38.5%) received it much less than expected, and (57.5%) were able to obtain social support

through opportunities to talk about financial matters, while (42.5%) did not receive it, and invitations to events represented social support. For (79%) of the sample, while it was not so for (21%) of the sample, social support was through useful advice in the form of (90%), while it was not the same for (10%). Social support for (84%), while it was not the same for (16%), and in the event of illness, assistance constituted a form of social support for (92%) and was not supportive for only (8%) of the sample.

Table 4.5: Distribution of the studied groups according to the quality of life results of thalassemia patients in Nasiriyah city, Iraq, with significant comparisons (n = 200)

Information on	**n (%)**	**Mean**	**± SD**	**Level of QOL**
I had enough energy for daily activities Never Rarely Occasıonally Mostly Always	 10(5%) 20(10%) 74(37%) 40(20%) 56(28%)	3.56	± 1.14	High
I found it hard to keep up physical activity with my peers Never Rarely Occasıonally Mostly Always	 28(14%) 24(12%) 70(35%) 18(9%) 60(30%)	3.29	± 1.37	Low
My health allows me to participate in as many social events as I would like Never Rarely Occasıonally Mostly Always	 26(13%) 24(12%) 42(21%) 26(13%) 82(41%)	3.57	± 1.44	High
Pain prevented me from doing what I need to do Never Rarely Occasıonally	 46(23%) 18(9%)	2.98	± 1.24	Low

Mostly Always	30(15%) 106(53%)			
I felt too tired to do the things I enjoy doing (i.e., hobbies, crafts, sports, musical instruments) Never Rarely Occasıonally Mostly Always	 30(15%) 18(9%) 70(35%) 18(9%) 64(32%)	3.34	± 1.39	Low
I was limited in my ability to do the kind of vigorous work I take part in (such as running, lifting heavy objects, participating in strenuous sports) Never Rarely Occasıonally Mostly Always	 22(11%) 12(6%) 66(33%) 10(5%) 90(45%)	3.67	± 1.38	Low
My treatments prevent me from doing the things I want to do Never Rarely Occasıonally Mostly Always	 110(55%) 30(15%) 14(7%) 20(10%) 26(13%)	2.11	± 1.47	High
I worried about getting an infection from the blood I receive Never Rarely Occasıonally Mostly Always	 66(33%) 16(8%) 30(15%) 12(6%) 76(38%)	3.08	± 1.72	Low
I worried about being able to have children Never Rarely Occasıonally Mostly	 98(49%) 20(10%) 30(15%) 20(10%)	2.34	± 1.54	High

Always	32(16%)			
I worried about being able to have children Never Rarely Occasıonally Mostly Always	 68(34%) 30(15%) 30(15%) 20(10%) 52(26%)	2.79	± 1.61	High
I worried thalassemia may lead to an earlier death Never Rarely Occasıonally Mostly Always	 114(57%) 20(10%) 20(10%) 16(8%) 30(15%)	2.64	± 1.69	High
I worried about painful tests Never Rarely Occasıonally Mostly Always	 86(43%) 18(9%) 30(15%) 14(7%) 52(26%)	2.64	± 1.67	High
I felt more anxious than other people in similar situations Never Rarely Occasıonally Mostly Always	 50(25%) 24(12%) 20(10%) 16(8%) 90(45%)	3.36	± 1.69	Low
I was bothered by my iron removal treatment Never Rarely Occasıonally Mostly Always	 30(15%) 20(10%) 40(20%) 20(10%) 90(45%)	3.60	± 1.50	Low
I was bothered by my transfusion Never Rarely Occasıonally Mostly Always	 38(19%) 16(8%) 40(20%) 14(7%) 92(46%)	5.30	± 1.57	Low
I felt angry more often				

than other people Never Rarely Occasıonally Mostly Always	 50(25%) 20(10%) 26(13%) 20(10%) 84(42%)	3.40	± 1.66	Low
I felt different from other people because of my disease complications and limitations Never Rarely Occasıonally Mostly Always	 54(27%) 12(6%) 36(18%) 16(8%) 82(41%)	3.30	± 1.66	Low
I felt sad Never Rarely Occasıonally Mostly Always	 39(19.5%) 13(6.5%) 40(20%) 20(10%) 88(44%)	3.52	± 1.56	Low
My family supports me and my health problems Never Rarely Occasıonally Mostly Always	 2(1%) 4(2.0%) 64(32%) 20(10%) 110(55%)	4.16	± 1.00	High
My friends support me and my health problems Never Rarely Occasıonally Mostly Always	 32(16%) 10(5%) 44(22%) 8(4%) 106(53%)	3.73	± 1.52	High
I was hopeful about my future Never Rarely Occasıonally Mostly Always	 26(13%) 12(6%) 44(22%) 20(10%) 98(49%)	3.76	± 1.43	High
My treatments posed a financial strain for me/my family (i.e., transportation		2.54	± ± 1.63	High

to/from hospital, equipment, time off work) Never Rarely Occasıonally Mostly Always	 88(44%) 20(10%) 36(18%) 8(4%) 48(24%)			
Thalassemia negatively affected my family Never Rarely Occasıonally Mostly Always	 110(55%) 10(5%) 12(6%) 68(34%) 0(0%)	2.19	± 1.39	High
I worried about paying for my medication/equipment Never Rarely Occasıonally Mostly Always	 30(15%) 10(5%) 34(17%) 20(10%) 106(53%)	3.81	± 1.49	Low
Thalassemia and its treatments affect my academic performance, or I did not attend school Never Rarely Occasıonally Mostly Always	 56(28%) 14(7%) 50(25%) 20(10%) 60(30%)	3.07	± 1.58	Low
I have had trouble keeping up with my work, or I do not work Never Rarely Occasıonally Mostly Always	 36(18%) 20(10%) 40(20%) 6(3%) 98(49%)	3.55	± 1.58	Low
I have had trouble keeping up with my schoolwork, or I did not attend school Never Rarely	 16(8%) 30(15%)	3.57	± 1.36	Low

Occasıonally Mostly Always	60(30%) 12(6%) 82(41%)			
Thalassemia limited my career opportunities Never Rarely Occasıonally Mostly Always	 38(19%) 40(20%) 30(15%) 8(4%) 84(42%)	3.30	± 1.61	Low
I am bothered because I missed school. or I did not leave school Never Rarely Occasıonally Mostly Always	 20(10%) 10(5%) 50(25%) 30(15%) 90(45%)	3.80	± 1.32	Low
I am satisfied with my sex life Never Rarely Occasıonally Mostly Always	 4(2%) 10(5%) 30(15%) 16(8%) 140(70%)	4.90	± 1.04	High

Table (4.4) indicates the Quality of life scores ranging from 0 to 100 were obtained and rated on two scales, high and low. For example, a score between 51 and 100 indicates a high quality of life, while 1-50 indicates a low quality of life, the percentage of patients who obtained a high quality of life was as follows; 56 patients (28%) had sufficient energy to do physical activities, 82 patients (41%) were allowed by health to continue physical activity with colleagues, 110 patients (55%) whose treatments did not prevent them from doing the things they wanted to do, 98 patients (49%) They were not worried about the ability to have children, 114 (57%) did not feel anxious about their early death, 86 patients (43%) did not feel anxious about the increase in their disease, 110 patients (55%) felt family support with health problems, 106 patients (53 %) Felt the support of friends in health problems, 44 patients (22%) were optimistic about their future, 110 patients (55%) their medicines did not exert financial pressure on their family, 110

patients (55%) did not affect their families negatively, 140 patients (70%) were convinced of their sexuality.

The percentage of patients who obtained a low quality of life was as follows: 70 patients (35%) had difficulty continuing physical activity with colleagues, 106 patients (53%) that pain prevented them from doing what they needed to do, and 70 patients (35%) felt very tired from Doing things they enjoy doing, 90 patients (45%) had limited ability to do the kind of work or activity they participate in such as running and lifting heavy objects, 76 patients (38%) were concerned about contracting an infection from the blood they received, 90 patients (45%) felt more anxious than the rest of the people in similar circumstances, 90 patients (45%) were bothered by the iron disposal medications, 92 patients (46%) were bothered by the blood transfusions, 84 patients (42%) felt more anger than the rest of the people due to complications from their disease 88 patients (44%) felt sad, 106 patients (53%) were worried about medication fees, 60 patients (30%) Thalassma affected their academic performance, 98 patients (49%) had difficulty keeping up with their work, 84 Thalassemia patients(42%) restricted their job opportunities, 90 patients (45%) were upset about missing their work.

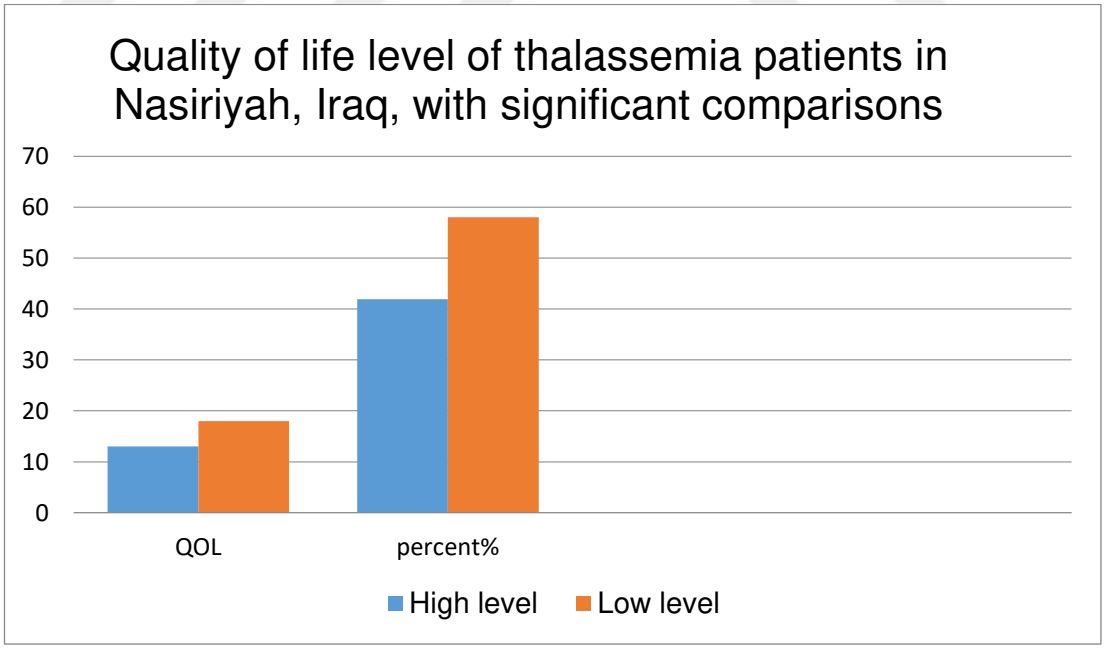

Figure 4.3: Quality of life level for thalassemia patients in Nasiriyah, Iraq

Table 4.6: Simple Pearson Correlation Coefficients between sociodemographic characteristics and quality of life

Demographical Characteristics X	Correlation	
Age	Pearson Correlation	1
	Sig. (1-tailed)	.000
	N	200
Gender	Pearson Correlation	.832**
	Sig. (1-tailed)	.000
	N	200
Mother Education	Pearson Correlation	.916**
	Sig. (1-tailed)	.000
	N	200
Father Education	Pearson Correlation	.883**
	Sig. (1-tailed)	.000
	N	200
Householder's	Pearson Correlation	.953**
	Sig. (1-tailed)	.000
	N	124
Family's Monthly İcome Iraq Diner	Pearson Correlation	.855**
	Sig. (1-tailed)	.343
	N	200
** Correlation is significant at the 0.01 level (1-tailed).		

Table (4.5) indicates the existence of a statistically significant correlation between demographic, economic and social data for thalassemia patients, and this relationship is positive.

5. Discussion

This chapter provides an explanation and discussion of the results of the study derived from the statistical analysis and its relevance to the objectives of the study, and further discussion will embody these objectives supported by the relevant literature and studies available.

5.1 Discussing the Demographics of Clients with Thalassemia in Nasiriyah

Table (4.1) The study indicats that the majority of the study sample (63%) are females and the rest were males. This study corresponds to the study by Majid et al., (2018) which concluded that the majority of the sample are females, so their percentage was (71.1%) and the rest were males, but these results differed with Kadhim et al., (2017) who showed that the majority (53%) are male and the rest are female, As for the age groups, most of them are between the ages of 20-18 years (40.5%). This is consistent with the study by Majid et al., (2018), as it shows that most of the ages are less than 25 years (42.7%), but it contradicts the study (Kadhim et al, 2017), which found To the majority of the sample from 6-15 years (41.7%)

As for the age groups, most of them are between the ages of 20-18 years (40.5%). this is consistent with the study by Majid et al., (2018), as it shows that most of the ages are less than 25 years (42.7%), but it contradicts the study (Kadhim et al, 2017), which found to the majority of the sample from 6-15 years (41.7%), as for the educational level, most of the study sample had obtained a sixth primary level (42%), and this corresponds to the study (Kadhim et al., 2017), they obtained the Elementary Shool, (30.40%), and it differed with the study by Majid et al., 2018), the majority of the sample obtained (44 %) on Medical field.

On the level of social support, most of them (84.5%) obtained social support through visits to friends and relatives, and the majority (81.3%) showed that, consistent with the study (Mikael and Al-Allawi, 2018), where the results of the study were that (68.98%) of the sample obtained Social support from relatives, and (59.26%) obtained emotional support from relatives, but it differs with the study by Al-Nuaimi et al., (2018) which shows that the majority of thalassemia patients (90.4%) of the sample obtained low social support.

5.2 Quality of Life for Clients with Thalassemia in Nasiriya

The results of the current study related to the quality of life of thalassemia patients were in agreement with the study conducted by (Adam, 2019). The study concluded that (46.6%) obtained a high quality of life, and it is consistent with the results of the study of (Al-Zahrani et al., 2017), which showed that thalassemia major patients who received Treatment in the past five years enjoy a good quality of life.

5.3 Discussion of the Correlation between Study Variables: Table (4.4)

Another important predictor of QOL was age (ANOVA analysis), QOL scores will decrease with increasing age as complications take effect and transition into adolescence and adulthood. Several other investigators have also documented such a decline in quality of life with age, including Ismail et al and Dahlawi et al. A study by Mikael and Al-Allawi, (2018), in which the study observed that Iraqi Kurdish children and adolescents with thalassemia had a decrease in QOL, affecting all domains. Be it physical or psychological.

6. Conclusions

The quality of life in Thalassemia patients in the Genetic Blood Diseases Center in the city of Nasiriyah was for more than half of the sample. It was mostly affected by the sample, the economic situation and social support, and the thalassemia disease limits the physical and emotional ability of the patients. This change can be explained by several factors, firstly. Above all, the use of iron medicines, which limited their physical abilities and thus reduced their quality of life, therefore all patients and their families (father and mother) must participate in health education programs to raise awareness about thalassemia disease and increase the participation of patients and their families in social networks and thus lead to improving the quality Life for the sick and increased social support from family and friends.

7. Recommendations

- Extending the investigation of this message to authorities and multidisciplinary health workers to conduct media campaigns.

- Involving patients and their families in educational sessions about the origin and development of the disease.

- Forming a technical team to guide patients to educate them on how to practice life with disease and participate in social and cultural activities, which increases their quality of life.

KAYNAKLAR

Adam, S., (2019). Quality of of life outcomes in thalassaemia patients in Saudi Arabia: a cross- sectional study, East Mediterr Health ;25(12):887–895, ***https://doi.org/10.26719/2019.25.12.887***

Alzahrani, R., Almutairi, O., Alghoraibi, M., Alabdulwahed, M., Abaalkhail, M., Alhawish, M., Alosaimy, M., (2017). Qality of life in transusion-dependent thalassemia patients, ***doi.org/10.1016/j.jtumed.2017.05.006***

Al-Nuaimi, M., Al- Hially, Y., Al- Hafidh, N., (2018). "β thalassemia major patients profile in Ninevah governorate-Iraq", ***Tikrit medical journal*** , 18(2): 250-260.

Amin, S., (2019). Everything you need to know about Thalassemia, healthline

Aljeesh, YI. (2016).Quality of Life among Thalassemia Children Patients in the Gaza Strip. American Journal of Nursing Science. Vol. 5, No. 3, 2016, pp. 106-113

Bajwa, Z, Basit, H., (2020). Thalassemia, NCBI**,** Treasure Island (FL): StatPearls Publishing

Borgna-Pignatti, C., Rugolotto, S., De Stefano, P., Zhao, H., Cappellini, M. D., Del Vecchio, G. C., Romeo, M. A., Forni, G. L., Gamberini, M. R., Ghilardi, R., Piga, A., & Cnaan, A. (2004). Survival and complications in patients with thalassemia major treated with transfusion and deferoxamine. Haematologica, 89: 1187–1193

Breda, L., Gambari, R., & Rivella, S. (2009). Gene therapy in thalassemia and hemoglobinopathies. Mediterranean Journal of Hematology and Infectious Diseases, 1(1), e2009008- e2009008.

Brissot, P; Cappellini, Maria D, (2014). "LIVER DISEASE". Thalassaemia International Federation.

Benz E, (2018)." Clincal Manifestatio and diagnosis of the thalassemias" , Waltham, Ma

Bethesda R, (2020), " HBB gene hemoglobin subunit beta" , Medline Plus Trusted Health information for you, U.S. National Library of Medicine

Cappellini, M. D. (2008). Long-term efficacy and safety of deferasirox. Blood reviews, 22, 3541.

Cappellini, M. D., Cohen, A., Piga, A., Bejaoui, M., Perrotta, S., Agaoglu, L& Capra, M. (2006). A phase 3 study of deferasirox (ICL670), a once-daily oral iron chelator, in patients with β thalassemia. Blood, 107(9), 3455-3462.

Cheerva, A. Crawford, J., Kutlar, A., Leonard, R., Raj, A.,(2020), Alpha Thalassemia Workup,Medscape, available at:https://emedicine.medscape.com/article/955496-workup

Dumars, K. W., Boehm, C., Eckman, J. R., Giardina, P. J., Lane, P. A., & Shafer, F. E. (1996). Practical guide to the diagnosis of thalassemia. American Journal of Medical Genetics, 62(1), 29 37

Daniel, E., and Sabath, MD., (2017). Molecular Diagnosis of Thalassemias and Hemoglobinopathies: An ACLPS Critical Review, *American Jornal of Clinical Pathology,* https://doi.org/10.1093/ajcp/aqx047

Dreuzy E, Bhukhai K, Leboulch PH, Payen E, (2016). " Current and Future alternative therpies for beta- thalassemia major" . *Biomedical Journal*, ScienceDirect,

Eiser C, Morse R, (2001). Quality of life measures in chronic diseases of childhood, Health Technology Assessment is indexed in Index Medicus/MEDLINE and Excerpta Medica/EMBASE. Copies of the Executive Summaries are available from the NCCHTA website (see opposite).;5(4)

Fessas, P. (1987). Prevention of thalassaemia and haemoglobin S syndromes in Greece. Acta haematologica, 78(2-3), 168-172.

Galanello, R., & Campus, S. (2009). Deferiprone chelation therapy for thalassemia major. Acta haematologica, 122(2-3), 155-164

Graziano, J. H., Piomelli, S., Hilgartner, M., Giardina, P., Karpatkin, M., Andrew, M., ... & Seaman, C. (1981). Chelation therapy in β-thalassemia major. III. The role of splenectomy in achieving iron balance. The Journal of Pediatrics, 99(5), 695-699.

Greer, John P.; Arber, Daniel A.; Glader, Bertil; List, Alan F.; Means, Robert T.; Paraskevas, Frixos; Rodgers, George M. (2013). Wintrobe's Clinical Hematology Lippincott Williams & Wilkins. ISBN 9781469846224.

Hashim, Z. A., & Mutlag, S. S. (2017). Ophthalmologic Changes in Thalassemia Patients. *Journal of Pharmaceutical Sciences and Research*, 9(11), 2237-2239.

Harewood, J., Azevedo. AM., (2020). Alpha Thalassemia, start pearls, available at https://www.ncbi.nlm.nih.gov/books/NBK441826/#_NBK441826_pubdet_.

Kadhim, K., Baldawi , K., Lami, F.,(2017), Prevalence, Incidence, Trend, and Complications of Thalassemia in Iraq" , international journal for hemoglobin research, ISSN: 0363-0269, ***doi:10.1080/03630269.2017.1354877***

Iawessome, (2012).,Alpha Thalassemia, available at: iawesome12.blogspot.com/2012/03/alpha-thalassemia.html?view=sidebar

Loukopoulos, D. (2011). Haemoglobinopathies in Greece: prevention programme over the past 35 years. The Indian Journal of Medical Research, 134(4), 572-572.

Palis, J., (2007) Diseases and disorders, " Pediatric Clincal Advisor Instant Diagnosis and Treatment",817-829, ISBN 978-0-323-03506-4, doi.org/10.1016/B978-0-323-03506-4.X1000-0

Porwit, A; McCullough, J; Erber, W. (2011). Blood and Bone Marrow Pathology. Elsevier Health Sciences. ISBN 978-0702045356.

Majid, M., Tutar, M., and Hashim, H., (2018). " Thalassemia awareness among Iraqi people in 2018, PagePress, Volume 10.8655,

Mettananda, S., Pathiraja, H., Peiris, R., Bandara, D., de Silva, U., Mettananda, C., & Premawardhena, A. (2019). Health related quality of life among children with transfusion dependent β-thalassaemia major and haemoglobin E β-thalassaemia in Sri Lanka: a case control study. Health and quality of life outcomes, 17(1), 1-13.

Mikael, N., and Al-Allawi, N., (2018). "Factors affecting quality of life in children and adolescents with thalassemia in Iraqi Kurdistan", US National Library of Medicine National Institutes of Health, 39(8): 799–807, ***doi: 10.15537/smj.2018.8.23315***

Musallam, K., Cappellini, M. D., & Taher, A. (2008). Challenges associated with prolonged survival of patients with thalassemia: transitioning from childhood to adulthood. Pediatrics, 121(5), e1426-e1429.

Muncie, Herbert L.; Campbell, James S. (2009). "Alpha and Beta Thalassemia". American Family Physician. 80 (4): 339–44. PMID 19678601

Origa, R., Bina, P., Agus, A., Crobu, G., Defraia, E., Dessì, C., & Galanello, R. (2005). Combined therapy with deferiprone and desferrioxamine in thalassemia major. Haematologica, 90(10), 1309-1314.

Origa, R. (2017). β-Thalassemia. Genet Med 19, 609–619

Pike, R., Bethesda, MD., (2016), Alpha thalassemia, MedlinePlus trusted health information for you available at: https://medlineplus.gov/genetics/condition/alpha-thalassemia/#references

Pinto V, Forni G, (2020). " Management of Iron Overload in Beta- Thalassemia Patients: Clincal Practice Update Based on Case Series" *International Jornal Molecular Sciences,* TDTInt. J. Mol. Sci. 2020, 21, 8771; doi:10.3390/ijms21228771

Pike, R., Bethesda, MD., (2017), Beta thalassemia, MedlinePlus trusted health information for you, U.S. Nastional Library of Medicine,8600 available at: https://medlineplus.gov/genetics/condition/beta-thalassemia/

Roy, T., & Chatterjee, S. C. (2007). The experiences of adolescents with thalassemia in West Bengal, India. Qual Health Res. 17 (1): 85-93.

Renshaw, T. L., & Cohen, A. S. (2014). Life satisfaction as a distinguishing indicator of college student functioning: Further validation of the two-continua model of mental health. Social Indicators Research, 117(1), 319-334.

Robin, E, Miller, MD., (2015) , Alpha Thalassemia, Kids Health for Parents, from *Nemours*, //kidshealth.org/en/parents/thalassemias.html?WT.ac=pairedLink

Rund, D. (2016). Thalassemia, modern medicine battles an ancient disease. *American Journal of Hematology*, 91(1), 15-21.

Rund, D., & Rachmilewitz, E. (2005). β-Thalassemia. New England Journal of Medicine, 353(11), 1135-1146.

Sabloff, Mitchell; Chandy, Mammen; Wang, Zhiwei; Logan, Brent R.; Ghavamzadeh, Ardeshir; Li, Chi-Kong; Irfan, Syed Mohammad; Bredeson, Christopher N.; Cowan, Morton J. (2011). "HLA-matched sibling bone marrow transplantation for β-thalassemia major". Blood. 117 (5): 1745–1750. doi:10.1182/blood-2010-09-306829. ISSN 0006-4971. PMC 3056598. PMID 21119108.

Shirzadfar, H., & Mokhtari, N. (2018). Critical review on thalassemia: types, symptoms and treatment. Advancements Bioequiv Bioavail, 1(2), 15-18.

Schwartz, M. (2012). The 5 Minute Pediatric Consult. Lippincott Williams & Wilkins. ISBN 9781451116564.

Steven, E, Mckenzie, MD.,(2011), Hematology, Pediatric Secrets, *Science Direct*, 302-336

Tamary H., and Dgany O., (2005), Alpha- Thalassemia, *NCBI*, University of Washington, Seattle

Thein S, (2013). " The Molecular Basis of B- Thalalssemia " , Cold Spring Harbor Perspectives in Medicine, **3(5): a011700, doi: 10.1101/cshperspect.a011700**

Torre, D, Lamb M, Geoffrey C.; Ruiswyk, Jerome V; Schapira, R M. (2009). Kochar's Clinical Medicine for Students. Lippincott Williams & Wilkins. ISBN 9780781766999.

Vichinsky, E., (2013), Clincal Manifestations of a- Thalassemia, Cold Spring Harb Perpect Med.; 3(5): a011742.doi: 10.1101/cshperspect.a011742

Verdugo M, Schalock R, Keith K, Stancliffe R, (2005). Quality of Life and its measurement important principles and guidleines, **49(Pt** 10):707-17, DOI : 10.1111/j.1365-2788.2005.00739.x

Weatherall, D. J. (2004). Thalassaemia: the long road from bedside to genome. Nature Reviews Genetics, 5(8), 625-631.

WHO, (1994), Measuring Quality of Life, available at: https://webcache.googleusercontent.com/search?q=cache:e0PNIoBnsUoJ:https://www.who.int/mental_health/media/68.pdf+&cd=1&hl=en&ct=clnk&gl=tr

WHO, (2006). WHOQOL-BREF. Research tools. [Online]. Available: https://www.who.int/substance_abuse/research_tools/whoqolb ref/en/

WHO, (2012), WHOQOL: Measuring Quality of Life, available at: https://www.who.int/tools/whoqol

WHO, (2018), Mental health: strengthening our response , available at:

https://www.who.int/news-room/fact-sheets/detail/mental-health-

strengthening-our-response

WHO, (2021).Achieving health for all,available at:
https://www.who.int/whr/1998/media_centre/executive_summary6/en/

Yu, U., Chen, L., Wang, X. et al. (2019). Evaluation of the vitamin D and biomedical statuses of young children with βthalassemia major at a single center in southern China. BMC Pediatr 19, 375

Zhou, Ya., Zhang, X., Liu, T., Wang, L., Yin, X., (2014). Splenectomy Improves Anaemia but does not reduce iron burden in patients with Haemoglobin H Constant Spring Disease, Blood Transfusion; 12(4): 471–478. doi: 10.2450/2014.0247-13**.**

Ek 1: Questionnaire

Al-Nasiriyah Şehrinde Talasemi'li (Akdeniz Anemisi olan) Hastalar İçin Yaşam Kalitesi

1. Yaş (Yıllar): 12-14 ( ) 15-17 ( ) 18-20 ( )

2. Cinsiyet: Erkek ( ) Kadın ( )

3. Medeni Durumu: Bekar ( ) Evli ( )

4. Eğitim Özellikleri (Yıl):

6^{th} ( ) 7^{th} ( ) 8^{th} ( ) 9^{th} ( ) 10^{th} ( ) 11^{th} ( ) 12^{th} ( )

Birinci Sınıf Öğrencisi ( ) İkinci Sınıf Öğrencisi ( )

5. Ailesinin ekonomik durumu:

5.1. Eğitim Seviyesi	Baba	Anne
Okuması ve yazması yok	( )	( )
Okur ve yazar	( )	( )
İlkokul	( )	( )
Ortaokul	( )	( )
Lise	( )	( )
Diploma	( )	( )
Lisans	( )	( )
Yüksek diploma	( )	( )
Yüksek lisans	( )	( )
Doktora derecesi	( )	( )

5.2. Hane Halkının Mesleği:

Meslek	
Profesyonel	( )
Yarı- Profesyonel	( )
Büro İşleri, Dükkan Sahibi, Çiftçi	( )
Vasıflı işçi	( )
Yarı Vasıflı İşçi	( )
Vasıfsız işçi	( )

İşsiz ( )

5.3. Ailesinin Aylık Geliri (Irak Dinarı):

< 300.000 ( ) 300.000-600.000 ( )

601.000-900.000 ( ) 901.000-1.200.000 ( )

1.201.000-1.500.000 ( ) ≥ 1.501.000 ( )

Sosyal Yardım

Duke-UNC İşlevsel Sosyal Destek Anketi

Başkalarının bizim için yaptığı veya bize yardımcı veya destekleyici olabilecek bazı şeylerin listesi aşağıda yer almaktadır. Lütfen her bir açıklamayı dikkatlice okuyun ve durumunuza en yakın boşluğa bir onay işareti (,) koyun.

Alıyorum ...	İstediğim kadar çok			İstediğimden az
Arkadaş ve akraba ziyaretleri				
Ev işlerine yardım				
Para yardımı				
İyi bir iş için övgü				
Önemseyen insanlar				
Sevgi ve ilgi				
Telefon görüşmeleri				
İş konuşma fırsatları				
Kişisel konuşma fırsatları				
Parayı konuşma fırsatları				
Davet				
Yararlı tavsiyeler				
Ulaşım konusunda yardım				
Hasta olduğumda yardım				

Transfüzyona (Kan Nakli) Bağlı Yaşam Kalitesi Anketi

	Hiçbir zaman	Nadiren	Bazen	Çoğunlukla	Her zaman
Günlük aktiviteler için yeterli enerjiye sahiptim					
Akranlarımla fiziksel aktiviteyi sürdürmekte zorlandım					
Sağlığım istediğim kadar sosyal etkinliğe katılmama izin veriyor					
Ağrı/sancı yapmam gereken şeyleri yapmama engel oldu					
Yapmaktan zevk aldığım şeyleri yapmak için kendimi çok yorgun hissettim (örneğin hobiler, el sanatları, spor, müzik aletleri gibi)					
Yer aldığım zorlu ve enerji isteyen işleri yapabilme yeteneğim sınırlandı (koşmak, ağır nesneleri kaldırmak, yorucu sporlara katılmak gibi)					
Tedavilerim yapmak istediğim şeyleri yapmama engel oluyor					
Aldığım kan nedeniyle enfeksiyon kapmaktan endişelendim					
Çocuk sahibi olabilme konusunda endişelendim					
Hastalık kapmaktan endişelendim					
Talasemi'nin daha erken ölüme yol açabileceğinden endişelendim					
Acı verici testlerden endişelendim					
Benzer durumlarda kendimi diğer insanlardan daha fazla endişeli hissettim					

Demirin atılması işlemimden rahatsız oldum					
Transfüzyonumdan rahatsız oldum					
Kendimi diğer insanlardan daha sık öfkeli hissettim					
Hastalık komplikasyonlarım ve kısıtlamalarım nedeniyle kendimi diğer insanlardan farklı hissettim					
Kendimi kederli hissettim					
	Hiçbir zaman	Nadiren	Bazen	Çoğunlukla	Her zaman
Ailem beni sağlık sorunlarım konusunda destekliyor					
Arkadaşlarım beni sağlık sorunlarım konusunda destekliyor					
Geleceğim hakkında umutluydum					
Tedavilerim benim/ailem için finansal bir yük oluşturdu (örneğin hastaneye / hastaneden ulaşım, ekipman, işten izin alma gibi)					
Talasemi ailemi olumsuz bir şekilde etkiledi					
İlaçlarımı/ekipmanımı ödeme konusunda endişeliyim					
Talasemi ve tedavileri akademik performansımı etkiliyor veya okula gitmedim					
İşimi sürdürmekte sorun yaşadım ya da çalışmıyorum					
Okul ödevimi takip etmekte zorlandım veya okula gitmedim					
Talasemi kariyer fırsatımı kısıtladı					
Okula gitmediğim için rahatsız oldum ya da okulu					

bırakmadım					
İşe gitmediğim için rahatsız oldum ya da işimi bırakmadım					
Seks hayatımdan memnunum					

Referans

Adib-Hajbaghery, M., Poormansouri, S. ve Ahmadi, M. (2017). Talasemiye Özgü Yaşam Kalitesi Anketinin Farsça Tercümesinin Psikometrik Değerlendirilmesi. *Hemşirelik Ölçüm Dergisi*, 25 (3), E202 – E217. https://doi-org.ezproxy.okcu.edu/10.1891/1061-3749.25.3.E202