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Long-Term Impact of Hemoglobinopathies upon Quality of Life and Behaviors of Adolescents in Blood Diseases Center in Karbala City.

Dissertation submitted By

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To the Council of College of Nursing, University of Babylon in partial fulfillment the requirement for the Degree of Doctorate of Philosophy in Nursing Sciences

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بِسْمِ اللَّـهِ الرَّحْمَـٰنِ الرَّحِيمِ

«وَنُنَزِّلُ مِنَ الْقُرْآنِ مَا هُوَ شِفَاءٌ وَرَحْمَةٌ
لِلْمُؤْمِنِينَ. وَلَا يَزِيدُ الظَّالِمِينَ إِلَّا خَسَارًا »

صدق الله العلي العظيم

(الآية ٨٢ من سورة الإسراء)

Dedication

- *To my father and my mother who support,help and encourage me to achieve the research project, without their love, help and support this project could not have been made possible.*
- *To My brother & my sister for supporting me with all love and respect.*
- *To My dear colleagues with all appreciation, and respect.*

Supervisor Certification

I *certify that this dissertation,* ***“Long- Term Impact of Hemoglobinopathies upon Quality of Life and Behaviors of Adolescents in Blood Diseases Center in Karbala City”*** was *prepared under my supervision at the College of Nursing, University of Babylon in partial fulfillment of the requirements for the Degree of Philosophy Doctorate in Nursing.*

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We, the examining committee, certify that we have read this dissertation entitled ***(Long- Term Impact of Hemoglobinopathies upon Quality of Life and Behaviors of Adolescents in Blood Diseases Center in Karbala City)*** which is submitted by the student (**Shahznan Hassan Badr)** from the Department of Pediatrics Nursing, and we have examined the student in all contents and we decide that it is adequate for awarding the **(Doctorate of Philosophy in Nursing)** with the specialty of Pediatrics Nursing.

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Abstract

Hemoglobinopathies are the most common life threatening, monogenic disorders in the world. The most common causes of hemoglobinopathies are sickle cell disease and thalassemia.

The main objective of the present study is to assess the quality of life and behaviours among Hemoglobinpathies adolescents with thalassemia and sickle cell anemia.

A cross-sectional descriptive study design was accomplished from 1st November /2020 to 3rd April/ 2022. The study involved (200) adolescent was carried out in Heredity Blood Diseases Center in Karbala Teaching Hospital for Children . The sample of the study was a non-probability convenient sampling technique.

The validity was determined through the distribution of the questionnaire to (13) experts. A pilot study was conducted on (20) adolescent to estimate the reliability of the questionnaire. The data were analysed through descriptive and inferential statistics.

The majority of adolescents are between the ages of 14 and 16. Female adolescents who graduate from primary school , had 1 sibling, and the second order within their family. Half of them study were regular therapy, and had the twice transfusion therapy. The highest percentage of parents were employed, most of parents graduate from institute and college graduated, Most of families were nuclear families, living in rural residents, and more than half slightly sufficient economic status. The negative health behaviors of less than half of adolescents were observed. Hemoglobinopathies adolescents' quality of life correlated

positively with the occupation of their father. Hemoglobinopathies adolescent quality of life and health behaviors showed a strong (positive) link.

The study concluded that there less than half of adolescent have poor health behavior and more than half of adolescent have moderate quality of life. As well as the quality of life and health behaviours of adolescents with hemoglobinopathies have been influenced by a variety of factors, including their parents' occupations, their mothers' education, and their families' socioeconomic status. In addition, the study concluded that there is significant effect of health behaviour on quality of life of hemoglobinopathies adolescents.

The study recommended that Adolescents with beta thalassemia and their mothers require comprehensive education should be maintained through a multidisciplinary approach over a prolonged outpatient, inpatient, and community education service plan for beta thalassemia Adolescents and their mothers.

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List of Abbreviations and Symbols

Abbreviations	**Full Terms**
%	Percentage
ABC	Adolescents Behaviors Checklist
ACS	Acute Chest Syndrome
CVD	Cerebra Vascular Disease
D.F	Degree of Freedom
F	Frequency
Hgb	Hemoglobin
HgbA	Hemoglobin Adult
HgbF	Hemoglobin Fetal
HgbS	Hemoglobin Sickle
HRQOL	Health Related Quality of Life
IV	Intravenous Infusion
M	Mean
PCA	Patient Controlled Analgesic
PQLI	Pediatric Quality of Life Inventory
PRBC	Packed Red Blood Cells
QOL	Quality Of Life
RBC	Red Blood Cells
SC	Sickle Cell
SCA	Sickle Cell Anemia
SCD	Sickle cell Disease
SD	Standard Deviation
Sig	Significant
SPSS	Statistical Package for the Social Science
TM	Thalassemia Major
WHO	World Health Organization

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Chapter One

Introduction

1.1 Introduction:

Hemoglobinopathies a collective term for severe inherited blood disorders are the most common life threatening, monogenic disorders in the world, with approximately 5% of the world’s populations are carriers for hemoglobinopathies . The most common causes of inherited hemolytic anemia are sickle cell disease (SCD) and thalassemia. SCD and thalassemia are inherited blood disorders, passed from parents to Adolescents through altered hemoglobin genes (Ahmed, 2018).

Thalassemia is caused by a decreased or absence of a hemoglobin (Hb) tetramer beta-globin chain synthesis consisting of two alpha and (2) beta-globin chains alpha2beta2. The clinical occurrence of beta-thalassemia is associated with the difference between chains of alpha globines and non-alpha globins (Hakeem et al., 2018).

Similarly, Hemoglobinopathies, according to Vicky at (2010), anemias caused by a structural or quantitative hemoglobin abnormality. These alterations might range from clinically insignificant to life-threatening. Sickling disorders, which are hemoglobinopathies, are hemoglobinopathies that cause red blood cells to take on a sickle shape under certain situations. One such example of a hemoglobin with structural abnormalities is hemoglobin S. Those with hemoglobin SS disease, the most common form of sickle cell anemia, are more likely to be deficient.

Hemoglobinopathy is a medical disorder characterized by the presence of hemoglobin that is abnormal. Fetal hemoglobin accounts for a significant portion of the newborn's hemoglobin supply (Hgb F). Hgb F levels decline in the first few months of birth as they are replaced by Hgb A (adult hemoglobin). In hemoglobinopathies, this normal hemoglobin configuration is disturbed. Causes of hemoglobinopathies are genetic and include sickle cell anemia, alpha-thalassemia, and beta thalassemia (Kyle and Carman, 2013).

Thalassemia is characterized by reduction or absent production of one or more of the globin chain that make up the hemoglobin (Hb) tetramers, which is the commonest autosomal recessive disorder worldwide. In general, thalassemia can be classified into two pivotal forms: α-thalassemia and β-Thalassemia. α-thalassemia is characterized by impaired synthesis of α globin chain and β- Thalassemia, the most common type of thalassemia, is due to impaired β globin chain synthesis. (Tahura, 2017).

In addition to , thalassemia is a complex heterogeneous group of diseases ranging from serious anemia to clinically asymptomatic individuals . It occurs due to diminution generation of hemoglobin. There are two fundamental sorts of thalassemia, Alpha thalassemia occurs when the imperfection occur in alpha globin gene while Beta thalassemia happened when the defect occur in beta globin gene .-Thalassemia is a condition in which the formation of -globin chains in the hemoglobin molecule is decreased or absent, resulting in an excess of -globin chains. The synthesis of -globin chains is decreased or missing in patients with -thalassemia (Odah et al., 2019).

In SCD, RBCs can become rigid and shaped like a sickle, and can clog small blood vessels, leading to oxygen failing to reach all parts of the body. This can result in tissue death and cause severe pain, as well as other complications, including overwhelming infections, lung problems and stroke. People with the SCD usually need long-life medical treatment (Ataga and Desai, 2018).

While, Pereira and others (2020) added that SCD is a monogenic disease resulting from a mutation in the gene responsible for the production of the hemoglobin A beta-globin, which determines changes in the resulting hemoglobin molecule (hemoglobin S).

Changes (variants) in the genes that provide information for creating globin chains generate changes in proteins, and this results in hemoglobinopathies. Depending on the specific mutation, globin chain levels may be lowered or altered, leading to structural changes in the globin chain. One of the hemoglobin chains (carrier) has a genetic variation that affects about 7% to 8% of the world's population, depending on ethnicity. Hemoglobin may be affected by genetic polymorphisms in the structure, behavior, production rate, and/or stability of the hemoglobin. In the presence of aberrant hemoglobin in RBCs, red blood cells' appearance and function can be altered. (Thom et al., 2013).

The word hemoglobinopathy is used to describe only those illnesses that have hemoglobin that is structurally defective (Hb). In other words, hemoglobin abnormalities of this kind are hemoglobinopathies. Diseases of hemoglobin production known as thalassemia are also known as quantitative hemoglobin disorders because they affect the production of one or more globin chains(CDC,2015).

The hemoglobinopathies are among the most widely recognized monogenic disease . Caused by gene mutations. In addition, ABO and Rh blood grouping are likewise important factor in transfusion and medicine and to reduce newborn hemolytic disease . Hemoglobinopathies status that influence kind and amount of hemoglobin .Hemoglobin is a tetramer protein made out of 2 sets of globin chains working in conjunction with 4 heme groups to transport oxygen in the blood (Mondal et al ., 2012)

Kohne (2011) was mentioned that the umbrella term hemoglobinopathy comprise all hereditary hemoglobin disorders .This may be either causing qualitative abnormalities as Sickle cell disease or causing quantitative abnormalities as Thalassemia .Sickle cell anemia, sickle cell disease, sickle cell syndrome, and sickle cell/-thalassemia are all autosomal recessive illnesses grouped together. Because of the replacement of amino acid valine for glutamic acid at the sixth position of the beta chain of hemoglobin, this genetic variation can be explained Sickle cell disease's pathogenic symptoms are related to the reduced lifespan of sickled blood cells (16-20 days compared to 120 days in normal red cells).

According to, the World Health Organization (WHO) who defines QOL as Individual Perception of their Position in Life in The Context of the Culture and Value System in Which They Live and Relation to Their Goals, Expectation Standard and Concern. Quality of Life Concept Is Affected by The Persons' Physical, Psychological State, Level of Independence, Social Relationship and Environment. Additionally, SCA Could Affect Quality of Adolescents's Life from All Dimensions. Physically Includes Diet, Physical Activity, Rest and Sleep, Management of Pain, Fever, And Vaso-Occlusive Crisis. Besides, Emotional Condition of Adolescents Could Also Be Affected Due to Body Image Disturbance, Fear, Anxiety About the Need for Future Treatment Can Interfere with Daily Living. Intellectual Aspect Also Can Be Affected as A Result of Learning Difficulties (Essawy et al., 2018).

Sickle cell-thalassemia are all autosomal recessive illnesses grouped together. Because of the replacement of amino acid valine for glutamic acid at the sixth position of the beta chain of hemoglobin, this genetic variation can be explained Sickle cell disease's pathogenic symptoms are related to the reduced lifespan of sickled blood cells (16-20 days compared to 120 days in normal red cells) (Kadri ,2019).

In individuals with SCD, the quality of life can be decreased by episodic and debilitating pain often associated with substantial analgesic use, frequent hospitalization and ultimately organ failure. HRQoL of adolescents with SCD may also be influenced by factors such as the inability to participate in peer group activities and the inability to keep up with developmental milestones (Adeyemo et al., 2015).

Patients' social, family, work, and recreational activities are negatively impacted by this disease's quality of life issues, which in turn raises the chance of hospitalization and mortality from this disease. These patients' quality of life and functional level, particularly their capacity to learn, were found to be closely linked, according to the researchers' findings (Madmoli et al., 2019).

Understanding the quality of life of adolescent is critical to evaluating and improving the care they receive. Chronic physical sickness, such as thalassemia, puts adolescent at risk for emotional and behavioral issues, which can have a negative impact on their life. When a adolescent is stigmatized as having the disease, it can lead to feelings of guilt and abandonment in the adolescent. It may also have a negative impact on social relationships, schoolwork, and self-confidence. As a result, throughout therapy and follow-up, the nurse must pay close attention (Eldakhakhny, 2011).

Adolescents with Thalassemia have more depressed symptoms and a lower quality of life than adolescent who suffer from short-term injury (QOL). Also, thalassemia adolescent are more likely to suffer from anxiety, despair, aggression, and shyness than their healthy counterparts, according to research. Furthermore, 47 percent of the youngsters had anxiety and depression, which affected their self-care and quality of life. The effects of thalassemia on a Adolescents's physical, emotional, and social development are all intertwined (Neshwan et al.,2018).

There are a number of factors related to the diseases and related to the social context that can affect quality of life. Ages, gender, level of education, the amount of symptoms and distress experienced by individuals with thalassemia have been related to quality of life (Eljedi,2019). In Iraqi especially in Kurdistan region the Adolescents and adolescents with thalassemia have a reduced HRQoL, affecting all domains whether physical, or psychosocial (Mikael and Al-Allawi, 2018).

Adolescence is a difficult developmental stage since it is a time when people are most open to new experiences and are more inclined to engage in dangerous behaviors that may have long-term consequences for their health. Chronic illnesses like SCD that necessitate hospitalization and long-term absences from school would increase the physical as well as other obstacles of this time (Meyiwa, 2019).

Adolescents are also characterized by the development of interpersonal interactions and peer support, according to Forman-Alberti (2015). As a way to fit in, adolescents with SCD may participate in risky behaviors such as smoking cigarettes or drinking alcoholic beverages. These can all contribute to feelings of sadness and low self-esteem, which can exacerbate the symptoms of the illness already present.

Accordingly, the feeling of failure and helplessness, somatization, impairment of self-esteem, and anger at being the victim of the unfairness of destiny, represent a supplementary affective burden for the adolescent and his family (Shaker and Nasir , 2013).

The average life expectancy and survival for individuals with SCD are influenced by physical, psychosocial, and environmental factors . Medical advances including penicillin prophylaxis, pneumococcal vaccines, blood transfusions for stroke prevention, the use of hydroxyurea, and stem cell transplantation have provided great benefits for individuals with SCD(Brewer, 2011).

1.2 Importance of Study

Thalassemia and sickle cell disease are important public health problems in Middle Eastern countries, including Iraq . These hemoglobinopathies are either due to reduction in the globin chain synthesis or to the production of abnormal chains (e.g. sickle cell disorders) . In Iraq, β-thalassemia is rather uniformly distributed throughout the country with an average carrier rate of around 4% and ∼ 15,000 registered patients with thalassemia major/intermedia . Sickle cell disorders, on the other hand, are less uniformly distributed with carrier rates varying from 0 to 16.0%, and they cluster in the extreme north and south of the country(Al-Allawi et al., 2015) .

The World Health Organization has reported that, Thalassemia has affected up to 270 million people globally, and that the Mediterranean region has 15 to 25 million individuals carrier (Tawfeeq, 2018). About 6-10% of the population in Iraq had hemoglobin apathy of which Thalassemia is a major part (Faraj et al., 2017).

While, Al-Hakeem and other (2020) pointed that There is only one center for recruiting, recording and treating all types of hereditary and non-hereditary hematological disorders called the Thalassemia Unit at the Al-Zahra hospital. Until the date of performing the current study, it contains 1022 records of patients who regularly visit the center for treatment and follow up. while, Jalal and others at (2009) stated that thalassemia major is an important health problem in Sulaimani province, with more than 600 registered cases in a population of over 1.5 million (Records of the Preventive Health Department- Sulaimani). Sulaimani is a large province lying in North Eastern Iraq, bordering Iran.

In addition to, Abdul-Zahra and others (2016) added that in Iraq, the carrier rate of b-TM in different governorates ranges between 3.7% and 6.5%.4 In Basra, the frequency of b-TM traits ranges from 3.3% to 7.9%, with an overall frequency of 4.6%.

According to World Health Organization(WHO), It Is Estimated That Each Year Over 300, 000 Adolescents with Severe Forms of SCA Are Born. The Majority of Them Are in Low and Middle Income Countries, about 2000 Adolescents Born with SCA Each Year in The United States. In Egypt at (2015), The Incidence of Sickle Cell Anemia Is 2,238 Adolescents. Whereas, In Iraq the Incidence Is 25,375 Adolescents at 2015(Aly Essawy et al., 2018).

As estimated by (Lee et al., 2019) that twenty-three thousands of newborn babies are diagnosed every year with major Beta-Thalassemia, compared with universal eighty million as a carrier. Almost, 56,000 babies every year born with severe β- or α-Thalassemia; of which in excess of half of them necessitate regular transfusion. Roughly (5,500) annual deaths during prenatal period; due to fetal hydrops, that result from the major α-Thalassemia.

Every year, between 50,000 and 100,000 Adolescents in low- and middle-income countries die of Thalassemia and about 7% of the world's population bears hemoglobin disorders (Alzahrani et al., 2017).

Sickle cell disease affects around 60-70 million individuals worldwide, with a prevalence of 7%. According to these figures, there are 15 million people with thalassemia and 240 million people who are beta-thalassemia carriers globally (2020, Dulhani and colleagues) .

For the adolescent with an infirmity or chronic illness, and for his family, there are exists specific problems in addition to those encountered by a healthy adolescent. The painful realization of social, professional and relational barriers is reactivated.in addition to, thalassemia Adolescents may require long-term psychological support. Cognitive-behavioral therapy can be an effective psychological approach because it contributes to treatment compliance, reduces emotional burden of disease and improves quality of life. (Shaker and Nasir, 2013).

Adolescents's physical, social, emotional, and educational functions are all negatively affected by thalassemia and its treatment, resulting in a lower quality of life overall. Feelings of hopelessness and depression have been the most typically reported symptoms, as well as difficulties with social integration and academic difficulties (Wahyuni et al., 2011).

The average life expectancy and survival for individuals with SCD are influenced by physical, psychosocial, and environmental factors. Medical advances including penicillin prophylaxis, pneumococcal vaccines, blood transfusions for stroke prevention, the use of hydroxyurea, and stem cell transplantation have provided great benefits for individuals with SCD (Oringanje et al., 2020)

Adolescence is a difficult developmental stage since it is a time when people are most open to new experiences and are more inclined to engage in dangerous behaviors that may have long-term consequences for their health. It would be even more difficult to deal with the physical and mental problems of this time if you had an illness like SCD, which requires frequent hospitalizations and protracted hospitalizations that lead to school absence (Meyiwa, 2019).

Peer support and the development of interpersonal interactions are hallmarks of the adolescent stage. Risky behaviors, such as smoking, drinking hard liquor, and having behaviors that can lead to depression and low self-esteem, may be undertaken by teenagers with SCD in attempt to fit in with their peers (Forman-Alberti, 2015).

In order to assess and enhance patient treatment, it is critical to gain a better grasp on what constitutes good health for Adolescents. Chronic physical sickness, such as thalassemia, puts Adolescents at risk for emotional and behavioral issues, which can have a negative impact on their life. When a Adolescents is stigmatized as having the disease, it can lead to feelings of guilt and abandonment in the Adolescents. It may also have a negative impact on social relationships, schoolwork, and self-confidence. As a result, throughout therapy and follow-up, the nurse must pay close attention (Eldakhakhny, 2011).

Mortality and morbidity due to hemoglobinopathies is a worldwide problem. Although they cannot be treated completely, genetic counseling and prenatal diagnosis can help prevent them. In the Mediterranean region, the Middle East, the Indian subcontinent, and other parts of South Asia, hemoglobinopathies (disorders of red blood cell formation) are quite common (Hussain et al., 2013).

Sickle cell disease affects around 60-70 million individuals worldwide, with a prevalence of 7%. According to estimates, 15 million individuals worldwide have thalassemia and 240 million people are beta-thalassemia carriers (heamog2). Every year, SCD affects an estimated 300,000 Adolescents around the world. It is anticipated that up to 80% of Adolescents with thalassemia will have mental health problems such oppositional defiant disorder, anxiety disorders, or depression. (Behdani et al., 2018).

While, Hassan and El Azzab (2016) reported that Its' global prevalence is about 2 per 1,000 newborns and 50,000 to 100,000

Adolescents die of thalassemia major in low and middle income countries while about 7% of the world's population is a carrier of a hemoglobin disorder . Thalassemia consists one of the most common chronic and genetic hematological disorder, globally. There are approximately 240 million people worldwide who are heterozygous for β-thalassemia while approximately 200,000 affected homozygotes are born annually.

The most severe form of -thalassemia, thalassemia major, results in acute anemia, according to Madmoli and others (2019). About 60,000 Adolescents are born each year with thalassemia around the world. Hotspots of this serious public health hazard include the Mediterranean region and the Middle East, as well as the Indian subcontinent and Southeast Asia's south and southeast.

Additionally, according to WHO that estimate 4-5% of the world’s population is carrier of hemoglobinopathies. Worldwide 15 million people have clinically apparent thalassemia disorders. With currently available medical treatment, affected adolescent have a low quality of life and a shortened life expectancy which leads to high mortality rate among the Adolescents (Mathew, 2018).

The hemoglobinopathies creates a burden not only on health system but also on the affected families, and Adolescents, which become vulnerable to emotional, social, psychological and behavioral problems (Hassan and El Azzab , 2016) . Patients with β-thalassemia suffering from disease chronicity including frequent hospitalization, blood dependence, continuous treatment with ICT; physical health limitations as growth retardation, poor physical appearance, and delayed puberty, in addition to disease complications (Tóthová et al., 2014).

While, Kaheni and others at (2013) added that states 44% of thalassemic patient have psychiatric problems, 74% have poor quality of life, 67% of patients have symptoms of anxiety, 62% have emotional problems especially depression, and 49% have communication problems.

Depending on the world health organization (WHO), Thalassemia is one of the greatest prevalent chronic genic disorders among 60 nations in the world, and it influence on 100000 adolescent activity around the world each year .The influences of thalassemia on physical wellness can lead to physical distort, growth blocking, and postponed maturity .The puzzles cited do not only impact patients' physical employment but also their emotional employment, social employment and school employment, conductive to weaken health-related quality of life (HRQOL) of the patients (Musa, 2017).

In comparison to patients who had short-term injuries, thalassemia Adolescents had more depressive symptoms and a lower quality of life, as mentioned by Burhan and others (2011). These findings emphasize the need for mental support and psychiatric and rehabilitation programs aimed at increasing motivation and quality of life for thalassemia adolescents. Evidence suggests that thalassemia patients have higher levels of anxiety, despair, hostility and shyness than healthy Adolescents. 9 There was another study that found that over half of the patients had anxiety and depression, which affected their self-care and quality of life.

1.3 Statement of the Problem

Long-Term Impact of Hemoglobinpathies upon Quality of Life and Behavior of Adolescents: Hemoglobinpathies include Thalassemia and Sickle Cell Anemia.

1.4.Objectives of the Study

1.4.1.General Objectives:

The major goal of this study is to evaluate the quality of life and behaviors of adolescents who have hemoglobinopathies.

1.5.2.Specific objectives

1. To assess the Quality of Life of Adolescents
2. To assess the Behaviors of Adolescents.
3. Determine the influence of Hemoglobinopathies on adolescents' quality of life and behavior.
4. To find out the relationship between the Quality of Life and demographic characteristics for Adolescents.
5. To find out the relationship between the Behaviors and demographic characteristics for Adolescent.

1.5 Research Questions

What are the Long-Term Impact of Hemoglobinpathies upon Quality of Life and Behaviour of Adolescents: Hemoglobinpathies include Thalassemia and Sickle Cell Anemia.

1.6 Research Hypothesis

Null hypothesis H0: there is no relationship between quality of life and behaviors of adolescent.

Alternative hypothesis H1: there is relationship between quality of life and behaviors of adolescent.

1.7.Definitions of Terms

1.7.1. Impact

a. Theoretical definition

Is an action of one object coming forcibly into contact with another (MacIntosh et al., 2017).

b. Operational definition

Refers to a physiological force of thalassemia or sickle cell anemia, an influence adolescents .

1.7.2. Hemoglobinpathies

a. Theoretical definition

It is the medical term for a group of blood disorders and diseases that affect red blood cells (Nubila et al., 2013).

b. Operational definition

Abnormal production or structure of the hemoglobin molecule such as thalassemia or sickle cell anemia caused by the abnormal hemoglobin that damages and deforms red blood cells.

1.7.3.Quality of Life

a. Theoretical definition

The standard of health, comfort, and happiness experienced by an individual or group (Wen et al., 2012).

b. Operational definition

A normal life without of thalassemia or sickle cell anemia experienced by an adolescents group.

1.7.4.Behaviours

a. Theoretical definition

A biological, cognitive, and social changes (Jaworska & MacQueen, 2015).

b. Operational definition

The way in which one acts or conducts oneself, especially towards thalassemia or sickle cell anemia.

1.7.5.Adolescents

a- Theoretical definition

Adolescent defined according the World Health Organization. Any individual between the ages of 10 and 19 (WHO., 2019).

b- Operational definition

Adolescence it is a period of transition in the development and growth from Adolescents to the adulthood, which ranges between (11-18years).

Chapter Two

Review of literature

2.1 Historical overview

First discovered by Thomas Cooley and Pear Lee in 1925, Cooley’ anemia or Mediterranean anemia is a genetic hematological condition. When Whipple and Bradford published their classic paper in 1932, they first used the term thalassemia to characterize the disease's pathophysiology. It is from the Greek word 'qalassa' that the name thalassemia was coined. Between 1925 and 1940, the first descriptions of thalassemia' clinical characteristics were published. Combining data from Europe and the United States between 1940 and 1960 provides a more complete picture of how the disease is passed down through generations. Between 1940 and 1950, the first detailed description of thalassemia's genealogical inheritance was made. According to Valentine and Neel (1944, 1948), the milder forms of cooley's anemia are referred to as thalassemia minor while the more severe variants are referred to as thalassemia major. Cooley's anemia sufferers' hemoglobin was shown to be more alkali-resistant by Vecchio in 1946 than that of healthy adults. Cooley's anemia patients may have higher levels of fetal hemoglobin than those who do not have the condition, according to the doctor's hypothesis. When cooley's disease causes microcytic anemia, big, pale macrocytes and target cells are frequently found along with the sickness. Target-cell anaemia, or leptocytosis, was first described by Dameshek, who observed this phenomena. Rich postulated in 1952 that thalassemia is caused by a lack of HbA synthesis and a continuous production of Hb F. For thalassemia, Ingram and Stretton's 1952 theoretical model is essential. The genetic regulation of thalassemia became apparent in the 1960s and 1980s. In 1970, Kan and Nathan reported on a moderate thalassemia intermedia sufferer whose parents both had HbA2 levels above normal, a condition known as intermedia (Weatherall and Clegg, 2008). Sephton-Smith proposed desferrioxamine, an iron cheltoe, as a groundbreaking treatment for thalassemia in (1962, 1964). In the 1980s and 1990s, thalassemia's molecular pathogenesis was finally unraveled (Nisha ,2013).

As a result, James B. Herrick (1910) was the first to define sickle cell anemia when he detected a sickling phenomenon in the blood of an African American student. As a result, his study, Peculiar Elongated and Sickle-Shaped Red Corpuscles in a Case of Severe Anemia, has been hailed as the first important work to deal with sickle cell anemia. Staphylococcus aureus (SCA) is a distinct disease entity transmitted by Mendelian law and mostly affects people of African descent, according to Huck Bugge (1923). It is possible that Emmel, in 1917, was the first person to use the word sickle cell while discussing the origins and creation of sickle-shaped erythrocytes. (Goddard and Gilmore, 1973).) Asymptomatic carriers of sickle cell disease are referred to as carriers, while sickle cell disease sufferers are referred to as carriers (Natarajan et al., 2010).

In addition, Walter Clement Noel, a Grenada dental student, was the first person to be diagnosed with Sickle cell disease in Chicago between 1904 and 1907. He died at the age of 37 because he was always sickly and pale. 15 Peculiar elongated sickle-shaped RBCs in the blood of an anemic black medical student were observed by James Herrick in 1910. In 1927, Hahn and Gillespie linked red blood cell sickling to low oxygen levels. Haemoglobin molecule structure alters when oxygen is not present, causing red blood cells to sickle. Watson hypothesized that the absence of illness symptoms in sickle cell neonates was due to the existence of fetal hemoglobin in the red blood cells (DAUDU, 2015). Using protein electrophoresis, Linus Pauling demonstrated in 1949 that sickle cell hemoglobin had a different structure than normal hemoglobin. A single amino acid substitution in the protein code causes sickle cell disease, according to research by Vernon Ingram and JA Hunt. After the Multicenter Study of Hydroxyurea in Sickle Cell Anemia was completed, Char ache claimed that the anticancer medication hydroxyurea was the first to minimize the frequent and unpleasant effects that define sickle cell sickness (DAUDU, 2015).

2.2 General overview of hemoglobiniopaties:

The hemoglobinopathies are autosomal recessive genetic disorders resulting in qualitative or quantitative change in the hemoglobin (Hb) molecule. HbS, the most widespread structural hemoglobin variant, hinders microcirculation by causing a sickle shape of red cells. In sickle cell disease, blood transfusion should be done only for strict indications. In thalassemia, Hb synthesis is impeded by defective production of globin chains. The commonest types of clinical importance are alpha and beta-thalassemia. Compound heterozygotes for thalassemia and Hb variants may have varying clinical impact(Politis, 2013) .

Hemoglobinopathies are a set of disorders that affect hemoglobin, which is the main structure of the red blood cells responsible for transporting oxygen throughout the body, and these disorders are lifelong and debilitating. These disorders are inherited and are considered genetic abnormalities, and they affect the structure and amount of the globin chain in the hemoglobin. There are thousands of types of hemoglobinopathies; some are silent, but the most common hemoglobinopathies are SCD and thalassemia, both of which follow the pattern of autosomal recessive inheritance (Kohne, 2011).

The incidence of hemoglobinopathies around the world pose a global health burden. Every year, an estimated 300,000 infants are born with inherited hemoglobinopathy disorders (of which 83% have sickle cell disease and 17% have thalassemia), with 80 percent of those Adolescents residing in countries with poor to medium incomes. According to Modell 27 and Darlison (2008), the global rate of hemoglobinopathies is 3.4 percent, and 7 percent of pregnant women around the world carry a Adolescents with a hemoglobin abnormality, which is the second most common cause of death in Adolescents under the age of five (Williams and Weatherall, 2012). 1 per cent of couples worldwide are at risk of having a Adolescents with hemoglobinopathies or already have a Adolescents diagnosed with a hemoglobinopathy; majority of these Adolescents die as a result of the condition (Alanazi, 2020).

The hemoglobinopathies, inherited disorders of the structure or synthesis of Hgb, are considered the common monogenic diseases. Approximately 80% of the annual births of babies with these conditions occur in low-or middle-income countries, many of which have extremely limited facilities for their control and management (Weatherall, 2013). Hgb disorders present a significant health problem in 71% of 229 countries (Salsabil et al., 2014).

Hemoglobin forms a tetramer with heme, which consists of two -globin and two non-globin chains, to transport oxygen throughout the body. Medical professionals refer to adult hemoglobin (HbA) as having an A 2A 2 chemistry. Hemoglobin variations are produced by combining the functions of the HBA1 and HBA2 -globin genes with those of the HBB structural genes (exons). A total of more than a thousand hemoglobin variations have been identified based on alterations in the globin chain. Hemoglobinopathies are caused by variations in the quality of amino acids. This condition is associated with quantitative changes such as amino acid insertion, deletion or mutation in the interstices (introns) and results in decreased globin chain production (Faried et al.,2019).

Worldwide, hemoglobinopathies are the most frequent monogenic disorders. Thalassemia syndromes and structural hemoglobin variations are two of the most common types (abnormal hemoglobin’s). Hemoglobin variations HbS, HbE, HbC and HbD are among the most common and clinically relevant structural hemoglobin variants in thalassemia. The cost of caring for patients is a significant burden on the global health system (De Sanctis et al., 2015).

The interaction between different thalassemia variants and the primary defective hemoglobin results in a wide spectrum of clinical diseases, all with various degrees of severity. Thalassemia major and intermedia are both possible in 1-3 people who are homozygous for -thalassemia. Thalassemia major is frequently discovered in the first two years of life and necessitates lifelong blood transfusions in order to maintain life expectancy. There is no need for blood transfusion in patients with thalassemia intermedia, which is identified later and usually results in milder anemia (Taher et al., 2021).

2.3. Categories of genetic disorders

Genetic disorders fall into four types. There are four main types of chromosome abnormalities: those caused by deletion, duplication, or modification of large segments of chromosomes, those caused by changes or mutations at the gene level, multifactorial disorders, and those caused by changes (mutations) in multiple genes and environmental factors, as well as mitochondrial-disorders (Thisen and Shaffer ,2010) .

2.4. Patterns of single-gene disorders

A single-gene condition is referred to as thalassemia or SCD. In 1865, Gregor Mendel, a monk, originally established four basic patterns for the transmission of single-gene diseases from parents to Adolescents. There are four distinct forms of inheritance: X-linked dominant, x-linked recessive, and autosomal dominant (Aimiuwu et al., 2012).

2.5. Thalassemia and sickle cell disorders

Thalassemia and SCD are single-gene illnesses that are passed down in families through the autosomal recessive mode of inheritance (Swensen et al., 2010). Males and females alike can be affected by a 'autosomal' disease since the anomaly is on one of the autosomal DNA. In order for a Adolescents to develop a severe clinical illness known as thalassemia major, they must both receive a faulty gene from both of their parents (Huang et al., 2020).

In the case of thalassemia offspring, the term heterozygote refers to a Adolescents who has a normal set of genes from one parent and a defective set from the other. Carrier is another term used to describe someone who has the thalassemia trait (thalassemia minor). These people will not experience any signs of the condition, but they could pass on the damaged gene to their Adolescents. Each pregnancy has a 25 percent (1:4) risk that the Adolescents will be a homozygote, meaning that the Adolescents will receive the faulty (thalassemia major) gene from both parents, i.e., a one in four probability (Langlois et al., 2008).

Thalassemia and SCDs are one of the world’s leading genetic conditions with over 5% of the world’s population carrying clinically significant Hgb gene variants. It is estimated that, each year, more than 330,000 live births are born with either thalassemia or SCD (17% thalassemia, 83% SCD) (Hoppe, 2013). Globally, Thalassemia and SCD are considered the highest prevalence of anemia in both sexes and all-time periods after iron deficiency anemia and hookworm. Anemia accounted for 68.4 million in 2010 (Chaparro and Suchdev, 2019).

2.5.1. Thalassemia

Hemoglobin anomaly known as Thalassemia is characterised by the absence or decrease of globin chains in blood. Malaria was found most frequently in tropical and subtropical regions of Mediterranean countries, the Middle East, Trans Caucasus, Central Asia, the Indian Subcontinent (South Asia), and Southeast Asia (Weatherall, 2006). -thalassemia and -thalassemia are the two most common kinds of thalassemia (Kotila, 2012).

Thalassemia is a frequent disease caused by mutations in the -globin gene locus, which results in lower levels of -globin production (Thein, 2013). Mutations that impact the expression of the -globin gene have been shown to produce -thalassemia in more than 200 different ways. Rather of being randomly distributed, these mutations have a distinct regional and ethnic origin due to the existence of a small number of common mutations mixed in with varying numbers of unusual ones (Cabras et al., 2011).

2.5.1.a. Definition and etiology of thalassemia

Disorders characterized by low or nonexistent levels of - or -globin production are collectively referred to as thalassemia. Unbalance is created when the rate at which one or more globin chains are synthesized alters (Shafique et al., 2021). Thalassemia as a hemoglobinopathy genetic condition of abnormal Hgb is inherited through an autosomal recessive pattern similar to those of SCD(Rahimi, 2013).

2.5.1.b. Incidence of thalassemia

Worldwide prevalence of genetic carriers of beta thalassemia is 3% . The thalassemia are found primarily in people of Mediterranean descent, although they have been reported in Asian and African populations. Thalassemia is on the rise among young Asian Adolescents in the United States (15). More people in 2015 were diagnosed with thalassemia than expected, with a prevalence rate of 34.6/100,000. (Hossain et al., 2017).

Thalassemia represented 75% of all hemoglobinopathies in Iraq and beta- thalassemia major represent 67% of all type of thalassemia (Ali and Abdulla, 2020)

2.5.1.c. Types of thalassemia

Different combinations of abnormal genes lead to more than 60 thalassemia syndromes. There are two basic types of thalassemia, α and β. The production of the Hgb protein's chain is impaired in -thalassemia. Alpha thalamic illnesses include Hgb H disease and Hgb Bart's hydrops fetalis (Tamary and Dgany, 2020).

Problems with the β chain occur more often and the condition β-thalassemia can be divided into three subcategories based on severity. Subcategories of thalassemia include, thalassemia minor (also called β-thalassemia trait) that leads to mild microcytic anemia; often no treatment is required, Thalassemia major (Cooley anemia) is the homozygous form of thalassemia intermedia, which necessitates frequent blood transfusions in the Adolescents (Karimi et al., 2012).

2.5.1.d. Pathophysiology

Hereditary blood illnesses known as the Thalassemia are characterized by a deficiency of adult hemoglobin production. This is because the RBCs are aberrant in both size and morphology, and they die quickly. Chronic anemia arises as a result of this conditional defect. The body tries to make up for the lack of oxygen by creating a lot of fetal hemoglobin. The normal synthesis of hemoglobin polypeptide chains is impaired in thalassemia, resulting in their occurrence. Thalassemia are caused. Greek letters identifying the polypeptide chain that is impacted are used to classify them, although the most prevalent are a and b thalassemia (Leifer, 2014).

Deficiencies in the beta globin gene are the primary cause of beta thalassemia. An inability to produce hemoglobin A's required beta globin chains results. Heterozygous individuals manufacture half as many beta globin chains as healthy individuals. For people who are beta thalassemia homozygotes, two subtypes exist: beta and beta0 (Bowden and Greenberg, 2010).

Those in the beta group can synthesize around one-third of the normal beta globin chains, but those in the beta0 group cannot. This does not alter the creation of alpha globin chains, which are essential for the synthesis of hemoglobin A, There are an excessive number of unlinked alpha globin chains since there are no beta chains to connect them. These free alpha globin chains precipitate and show as inclusion bodies in the growth of red blood cells. As a result of the inclusion bodies' toxic effects, the majority of bone marrow's red blood cells die. Red blood cells that reach the peripheral circulation in lower quantities are characterized by microcytosis (small size), hypochromic (reduced color as a result of poor staining qualities due to reduced hemoglobin), poikilocytosis (abnormal shape), and the presence of inclusion bodies (Kwaifa et al., 2020).

The spleen and liver rapidly remove damaged cells from circulation, thereby reducing their lifespan. Red blood cells with a higher concentration of hemoglobin F have a better chance of surviving the body. Hemoglobin F may be produced by thalassemia patients in order to compensate for the absence of hemoglobin A. When the bone marrow swells as a result of severe anemia, an increase in the generation of erythroid precursor cells occurs. The increased erythropoiesis is balanced out by hemolysis in these cells, much like their ancestors (Petrakos et al., 2016).

2.5.1.e. Clinical manifestation of hemoglobinpathies disorder

The onset of clinical manifestations in thalassemia major may be insidious and not recognized until late infancy or early toddlerhood. Bony cortex thins and the bone marrow expands as a result of the increased activity. Growth retardation, pathologic fractures, skeletal deformities (frontal and maxillary bossing), pubertal delay, severe anemia, characteristic faces (enlarged head, frontal and parietal bossing, severe maxillary hyperplasia, malocclusion), hepatosplenomegaly and a bronze skin tone (Hussein, 2019).

Anemia, fatigue, pallor, irritability, and anorexia are noted in Adolescents with thalassemia. Bone pain and fractures are seen. Many body systems can be affected, including enlargement of the spleen, overstimulation of bone marrow, and heart failure. The liver, gallbladder, and pancreas can also be involved. The skin may appear bronze-colored or jaundiced. Skeletal changes occur, including deformities of the face and skull. The upper teeth protrude, the nose is broad and flat, and the eyes are slanted (Hatfield, 2007).

2.5.1.f. Diagnostic tests of hemoglobinpathies disorder

An individual's symptoms, medical history, and test results all go into making a thalassemia diagnosis. Thalassemia can be detected through specific newborn screening tests, which are collected according to state law. Prenatal diagnosis is also possible. One of the most striking laboratory findings is the extremely low hematocrit, which can fall to less than 10% of normal if not treated with transfusions. Thalassemia minor, however, often presents with an anemia so mild, it is mistaken for iron-deficiency anemia (Nicki et al., 2012).

Hematologic studies reveal characteristic changes in the RBCs (e.g., microcytosis, hypochromic, anisocytosis, poikilocytosis, target cells, and basophilic stippling of various stages). Low hemoglobin and hematocrit levels often occur in severe anemia, although they are typically less pronounced than the reduction in the RBC count because of the proliferation of immature erythrocytes. In order to determine the kind and severity of thalassemia, hemoglobin electrophoresis is used to determine the quantity and type of hemoglobin variations present in the patient's blood. Because neither Hgb F nor HgbA2 (a kind of normal adult hemoglobin) is synthesized using -chain polypeptides, their levels are high in people with -thalassemia (Hockenberry and Wilson, 2018).

2.5.1.g. Medical Management

In order to prevent bone marrow expansion and bone abnormalities, the goal of supportive therapy is to keep hemoglobin levels high enough to allow growth and regular physical activity. The goal of medical care is to keep the hemoglobin level over 9.5 g/dl, which may necessitate transfusions up to three times a month.. Its benefits include enhanced physical and psychological well-being, less cardiomegaly and hepatosplenomegaly, fewer bone alterations, normal or almost normal growth and development till adolescence, and fewer infections (Perry et al., 2017).

The medical management of beta-thalassemia is based on three main points: monitoring Hgb, hematocrit, erythrocyte transfusions, monitoring blood iron levels, providing chelation therapy, and splenectomy. To prevent the severe side effects and bony changes associated with the disease, the Hgb is maintained at approximately 10 mg/dL, although this parameter is often individualized (Abdulaal, 2018 ; Chonat and Quinn, 2017)

2.5.1.g.1-Blood transfusion:

Anemia is corrected, erythropoiesis is suppressed, and gastrointestinal iron absorption is inhibited as a result of enhanced, but inefficient, erythropoiesis in non-transfused patients. Anemia that has persisted for more than two weeks, with no other contributing factors such as infections, should be used as the basis for starting transfusions in patients with a confirmed diagnosis of thalassemia (Langhi et al., 2016). Patients with Hb greater than 7 g/dl should also be evaluated for facial changes and signs of bone enlargement as well as growing splenomegaly. Regular transfusions should not be started until the second or third year, if at all possible, due to the possibility of developing numerous anti-red cell antibodies and the ensuing difficulties in obtaining acceptable blood donors (Lal, 2020). Most commonly, however, a pre transfusion hemoglobin level of 9 to 10 g/dl is recommended, followed by an after transfusion hemoglobin level of 13 to 14 g/dl. As a result, Adolescents's growth, organ damage, an4d bone deformities are protected, allowing them to lead normal, productive lives (De Sanctis et al., 2013). The frequency of transfusion is usually every two to four weeks. Shorter intervals might further reduce the overall blood requirement, but are incompatible with an acceptable quality of life. The amount of blood to be transfused depends on several factors including weight of the patient, target increase in Hb level and hematocrit of blood unit. Appropriate graphs and formulae to calculate the amount of blood to be transfused are available (Ikehara, 2011).

Chronic anemia can be alleviated by blood transfusions that prevent bone abnormalities as well as allowing for a healthy quality of life for the patient (QoL) Anemia is corrected and erythropoiesis is suppressed by RBC transfusions, which prevents hepatosplenomegaly and limits bone marrow hyperplasia by providing fresh, normal RBCs (Shah et al., 2019).

Packed red blood cells for patients with -thalassemia should have a total Hb concentration of at least 40 g. The blood should be obtained from no remunerated donors, processed, tested, stored, and supplied by blood transfusion centers of the highest quality. In the presence of anemia and/or clinical symptoms, such as failure to thrive (identified as reduced growth velocity and delayed developmental milestones), decreased QoL, and comorbidities such as organ dysfunction and extra medullary hematopoiesis, the decision to begin blood transfusions is appropriate. A criterion of 70 g/L (tested on two occasions approximately two weeks apart, eliminating other reasons for the anemia) or the presence of clinical problems regardless of Hb level is recommended by TIF, UK, and US guidelines for blood transfusions (Cappellini, 2008).

2.5.1. g.2. Chelation Therapy

An iron balance can be achieved with chelation therapy by increasing iron excretion through urine and/or feces. The rate at which iron must be excreted will need to be higher if chelation has been delayed or inadequate. Chelation therapy's main problem is to strike a balance between its beneficial effects and the undesirable side effects of excessive chelation, because iron is necessary for critical physiological tasks. To avoid excessive chelation as iron levels drop, careful dose modification is required (Azman, 2017).

In chelation therapy, the second key problem is to maintain regular treatment regimens throughout one's life, as even short periods of treatment interruptions can have detrimental effects. As significant as individual chelates are, other factors, such as psychological well-being, family and institutional support have an impact on adherence and outcomes as well (Cappellini et al., 2014; Poggiali et al., 2012).

In treating thalassemia, the most common issue is an excess of iron in the body. Iron chelates are the sole option to get rid of extra iron because the body lacks effective mechanisms for excreting it. There is a primary goal of using iron chelates to reduce iron storage in the body and to maintain a low amount of iron in the body. Desferroxamine, Deferiprone, and Deferasirox are among the current medications in use (Kontoghiorghe et al., 2016).

For six nights a week, a subcutaneous infusion pump administers 30-40mg/kg/day of desferrioxamine subcutaneously over eight to ten hours. Desferrioxamine in depot form is a more recent development in chelation therapy.

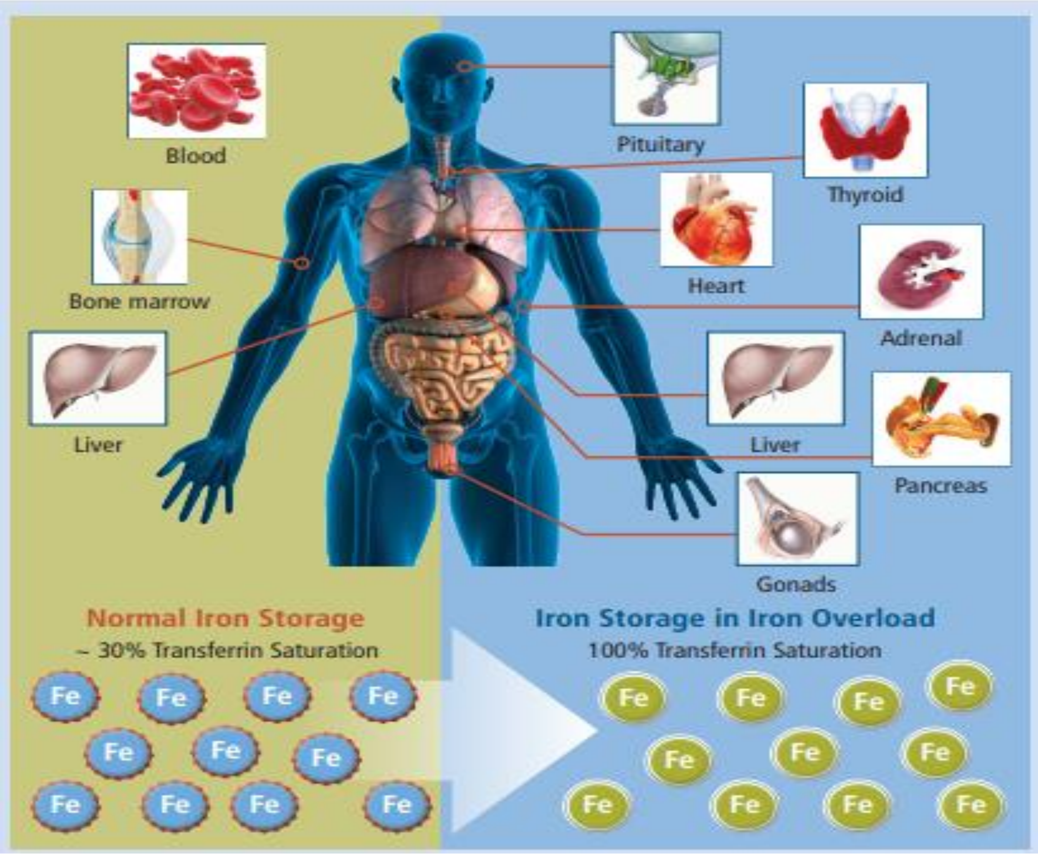
Three to four split dosages of 75-100mg/kg/day are taken orally three to four times a day. In comparison to desferrioxamine, it has been proven to be 70 to 100 percent as effective in reducing both serum ferritin and tissue iron overload (Manglani, 2012).

If you've had a lot of blood transfusions, you may need to take this new oral iron chelator, Deferasirox. The dose is 20-40mg once daily, and it is changed based on the patient's response, ferritin and creatinine levels in the blood. In animal trials, it was found to be ten times more potent than deferiprone and nearly five times as efficacious as subcutaneous desferrioxamine (Fisher et al., 2013).

2.5.1. g.2.1. Assessment of iron overload

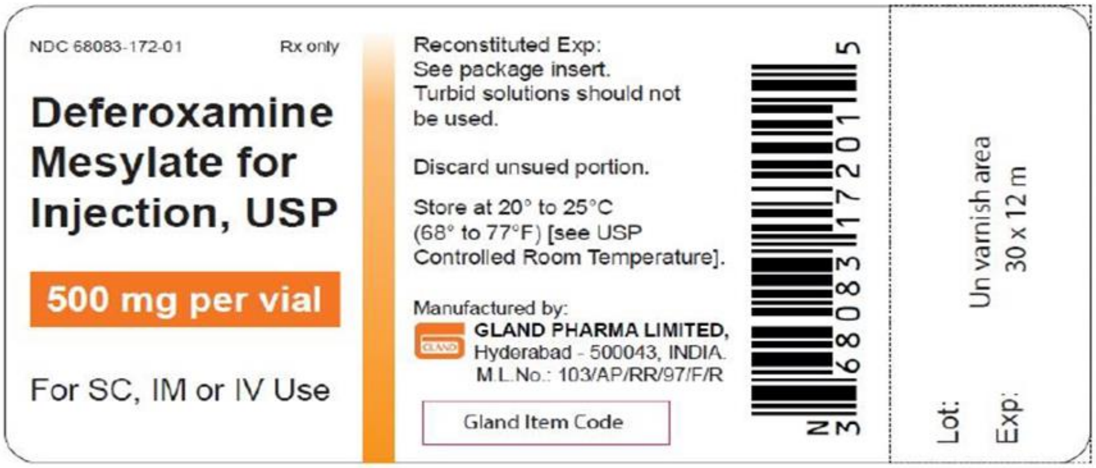
Dietary iron is absorbed from the gut and circulates in the blood bound to the protein transferring (Figure1). Three main methods for perfect assessment of transfusion iron loading are; net volume of blood infused per year, serum ferritin, and liver iron concentration. To determine the daily rate of iron loading, it is necessary to keep track of the total volume of red blood cells infused every year (ml/kg red cells) at each transfusion visit (Quinn and St Pierre, 2016).

Iron chelation therapy



Source: **Carson, S. M., and Martin, M. B. (2014).** Effective iron chelation practice for patients with β-thalassemia major. *Clinical journal of oncology nursing*, 18(1), 102-111.

Desferrioxamine needs to be given most effectively by subcutaneous infusion pump, to avoid hospitalizing Adolescents who require deferoxamine therapy, between 20 and 60 mg/kg of body weight throughout an 8- to 12-hour period, three or six times a week, typically (Campos,2014). Chelation is a viable alternative for motivated youngsters and their parents, as long as they and their caregivers adhere to the regimen. Long bone and spine development plates can be damaged, hearing loss and retinopathy can occur, and these are the most prevalent adverse effects in Adolescents. It is possible to avoid these adverse effects by using a dose of 20–40 mg/kg alone (Uppuluri and Raj, 2021).



NDC 68083-172-01 Rx only

Deferoxamine Mesylate for Injection, USP

500 mg per vial

For SC, IM or IV Use

Reconstituted Exp:
See package insert.
Turbid solutions should not be used.

Discard unsued portion.

Store at 20° to 25°C
(68° to 77°F) [see USP Controlled Room Temperature].

Manufactured by:
GLAND PHARMA LIMITED,
Hydorabad - 500043, INDIA.
M.L.No.: 103/AP/RR/97/F/R

Gland Item Code

N 3 68083 17201 5

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30 x 12 m

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Source: Deferoxamine - FDA prescribing information, side effects and uses. (n.d.). Retrieved February 24, 2018, from https://www.drugs.com /pro/deferoxamine.html

Deferasirox (Exjade), an oral chelation agent, is now available for Adolescents 2 years of age and older and is taken on an empty stomach at least 30 minutes before each meal. In order to take Deferasirox, you must dissolve it in water, orange or apple juice. Compliance is improved with this orally administered medication. However, it is not without serious side effects, including gastrointestinal hemorrhage, Adolescentsney and liver damage (Shah,2017).

Drug DFP is currently licensed to treat iron chelation. Doses of 25 mg/kg three times a day might be raised depending on the Adolescents's reaction and treatment goals. There is a maximum daily intake of 33 mg/kg of body weight. The major adverse reactions of DFP are agranulocytosis and arthroplasty. Obtain an absolute neutrophil count before starting deferiprone therapy and then weekly thereafter, as ordered, because drug can cause agranulocytosis that can become life-threatening (Saliba et al., 2015).

Deferiprone

Manufactured by: Apotex Inc., Toronto, Ontario, Canada, M9L 1T9

Distributed by: ApoPharma USA, Inc. Rockville, MD 20850

223190

NDC: 52609-0006-1

Rx only

Ferriprox®

(Deferiprone) Tablets

500 mg

Attention Pharmacist: Dispense the accompanying Medication Guide to each patient.

APO 500

100 Tablets

Each film-coated tablet contains: 500 mg deferiprone.

Usual Dosage: See the package leaflet for full prescribing information.

Keep out of the reach and sight of children.

Store at 20° to 25°C (68° to 77°F); excursions permitted to 15° to 30°C (59° to 86°F) [See USP Controlled Room Temperature.]

3 52609 00061 6

Pull open

ApoPharma

Source: Ferriprox. (n.d.). Retrieved February 24, 2018, from https://www.drugs. com/uk/ferriprox.

Thalassemia major patients should begin chelation therapy once they have received 10 to 20 transfusions or when serum ferritin levels climb to more than 1000 g/dl55, according to standard practice. Deferiprone monotherapy has been proven to minimize myocardial sidereosis and cardiac morbidity and death in prospective and retrospective studies48,49,50. In the early phases of iron overload, intensive therapy with iron chelates appears to improve glucose tolerance, aberrant thyroid function, and other adverse consequences (Bayanzay and Alzoebie, 2016).

The goal of iron chelating therapy to reduce the iron burden; secondly, to diminish the risk of tissue injury, particularly in particular main organs such as the heart and liver; thirdly, to increase the longevity of life;

fourthly, to provide protection along 24-hour against the iron toxic effects such as Labile Plasma Iron; and eventually, to decrease the free distance of iron chelating drugs (Mahmood et al., 2014).

Thalassemia's patients require blood transfusions and are mostly overloaded with iron unless they are undergoing iron chelation because their bodies are cannot excrete unnecessary iron. For these transfusion-dependent patients, it is important to choose an effective iron chelator to reduce their iron strain and enhance their quality and length of life. Desferrioxamine (DFO), deferasirox (DFX), and deferiprone (DFP) are the major iron chelates commonly used in clinical use. Numerous management methods have been suggested for decreasing overload of iron using these compounds. DFO treatment has been used as the standard gold for many decades. However, DFO is not absorbed orally and demands constant sluggish infusion, resulting in poor patient compliance, which adversely affecting its effectiveness (Lin et al., 2019).

While adequate medical treatment has minimized patients' difficulties, the psychosocial challenges they encounter are also of primary importance (Baiee et al., 2015).

2.5.1.g.3 Splenectomy

Splenectomy may be indicated in Adolescents who are requiring 200 to 250 mL/kg of packed RBCs each year to maintain their Hgb at 10 g/dL; the result is a moderate reduction in their transfusion requirements to 200 mL/kg per year. Adolescents after splenectomy are susceptible to infection (such as neisseria meningitidis, streptococcus pneumoniae, and hemophilus influenza) particularly in those younger than 5 years. Standard therapy for asplenic Adolescents includes immunizations, prophylactic penicillin, and a high index of suspicion and aggressive antibiotic therapy for febrile illnesses (Chonat and Quinn, 2017).

2.5.1.g.3.1. Indications for splenectomy:

A packed cell transfusion is required when the yearly demand exceeds twice the baseline value. i.e. about 230-250 mL per kilogram. Having a lower platelet count is an early sign of hypersplenism. Pneumococcal, H. influenza type b, and meningococcal infections should be inoculated six to eight weeks prior to splenectomy. Splenectomy should be avoided in Adolescents under the age of five (Iolascon et al., 2017).

2.5.1.g.4. Bone marrow transplantation

Currently, the only treatment option for thalassemia is a bone marrow transplant. According to the European and United States transplant registries, more than 1,200 Adolescents have received transplants from Hatched sibling donors. There are only 5% fatalities, 5% rejections, and 90% cures when a matched sibling donor is used with a myeloablative regime. Thousands of transplants have been a success. Even for Adolescents with a compatible donor and modest risk factors, only a tiny number (30%) can benefit from this treatment (Lucarelli et al., 2012).

When a Adolescents has a history of ischemic stroke or severe cerebral vasculopathy or has abnormal transcranial doppler flow with evidence of ischemic cerebral damage or multiple red cell alloantibodies that limit the availability of a sibling donor, the British Bone Marrow Transplant Society (BBMTS) says that sibling HSCT may be indicated (e.g. likely return to home country where health care services are inadequate). Long-term negative effects of HSCT include decreased fertility due to primary gonadal failure, skeletal issues, and a very low risk of malignancy (Oringanje et al., 2020).

2.5.1.g.5. Future gene therapy

The clinical applications of next generation sequencing of both DNA and andribonucleic acid (RNA) have become applicable to a wide range of clinical disorders. The important value of gene therapy is its clinical potential for the more effective prevention and management of single gene disorders, particularly the different forms of hemoglobinopathies. Understanding the basis of genetic is vital for the early assessment of hemoglobinopathies prognosis and future management. The role of recent genome-wide association to identifying genes that modify the level of Hgb F in β-thalassemia (Aldemir, 2018).

2.5.1.h. Complications of thalassemia

Hemostasis is another repercussion (excessive iron supply). hemolysis of RBCs and decreased Hgb production due to severe anemia improve iron absorption. Bronze coloring of skin, bone alterations and altered organ function are all caused by the excess iron that accumulates in the body's tissues. Osteoporosis, liver and gallbladder disorders as well as leg ulcers are among the other side effects of splenomegaly (Ricci and Kyle, 2009).

Many of the complications of thalassemia seen in developed countries today are the result of iron overload: 1. Liver: fibrosis and cirrhosis, 2. Beta cells of the pancreas: diabetes mellitus, 3. Pituitary, testis, and ovaries: growth retardation and hypogonadotropic hypogonadism, 4.

Parathyroid: hypocalcaemia, 5. Heart: arrhythmias, myocarditis, and intractable cardiac failure, Most, if not all, of these complications can be avoided by the consistent use of an iron chelator (Kurtoglu et al., 2012).

Thalassemia major patients can suffer a wide range of symptoms, some of which are minor while others are potentially life-threatening. These concerns can develop at any time in a patient's life. Chelating therapy and keeping blood hemoglobin levels within the normal range for the patient's age are often linked to difficulties in the majority of cases. 68 Adolescentshood treatment programs contain these two factors as well (Sadullah et al., 2020). BTM has a number of issues, including the following:

2. systemic bleeding and increased susceptibility to infection after blood transfusions or chelating therapy 5. Osteoporosis, a condition spread through blood, such as the hepatitis B virus, which causes bone loss.

Dysrhythmias of the heart

Delay in the onset of puberty is one example of a growth defect.

Splenectomy and hypothyroidism are examples of liver and hormone problems. Different thalassemia kinds may have an effect on a particular patient's well-being (Taher and Saliba, 2017).

When it comes to anemia, for example, the severity varies from patient to patient. This can vary from mild to severe in some Adolescents, depending on the degree of the anemia. Many factors affect a Adolescents's health and well-being and the occurrence of difficulties, including the need for many blood transfusions and the effectiveness of chelating therapy in cases of BTM. Also known as splenomegaly and "bone marrow abnormalities, these symptoms are common in patients with Thalassemia. When iron builds up in the heart and liver, it affects the endocrine system and other organs. As a result, some patients' social lives may be adversely affected by these issues (Quinn, 2013).

In these circumstances, individuals are more likely to suffer from bone deformities, diabetes, organ failure, or endocrine system abnormalities. Patients and their families could have been adversely affected by these issues. According to previous research, osteoporosis is a prevalent side effect even in BTM patients who have received adequate treatment. Osteoporosis is the most common consequence, followed by hormonal abnormalities (Boniel et al., 2021).

2.5.1.i. Nursing management

nurses' primary role in the care of adolescent who suffer from beta-thalassemia involves encouraging compliance with transfusion and chelation therapy, assisting Adolescents and families in dealing with the anxiety-provoking treatments and effects of the illness, and keeping an eye out for complications resulting from multiple transfusions (El-Said Zaghamir et al., 2019; Hockenberry and Wilson, 2013).

The nursing responsibilities outlined in general guidelines that apply to all transfusions; monitor the vital signs before administering blood then 15 minutes after initiation, then hourly, and on completion of the transfusion, check the blood group of the recipient against the donor’s, administer the first 50 ml of blood or initial 20% of volume (whichever is smaller) slowly and stay with the Adolescents, administer blood with normal through an appropriate filter, use blood within 30 minutes of its arrival from the blood bank and if it is not used return it to the blood bank; do not store it in a regular unit refrigerator, infuse a unit of blood within 4 hours, if a reaction of any type is suspected, take vital signs, stop the transfusion, maintain a patent IV line with normal saline and new tubing, notify the practitioner, and do not restart the transfusion until the Adolescents’s condition has been medically evaluated (AL-Naely et al., 2021).

Medication administration in nursing programs is a skill that nursing students must master prior to entering the nursing workforce. Nurse experts must check and encourage medication adherence at least once a month when Adolescents get transfusions or are visited at home. Adherence issues can be caused by intolerance, adverse effects, and lack of motivation (particularly when DFO injections are required on a regular basis). When it comes to improving patient adherence, the nurse specialist's job is critical (Hanson and Hadfather, 2021).

The nurse vital role before deferoxamine administration concluded in monitoring injection site and frequent moving infusion to another site, monitoring intake and output ratio, reporting any change or blood observing in stools especially at conditions of low platelet counts, and monitoring lab tests particularly baseline and periodic Adolescentsney function tests, serum ferritin, and platelet counts (Cappellini et al., 2014; Brown and Cutler, 2012)

Nurse having important role to educate Adolescents and family about deferoxamine chelate makes urine turn a reddish color, report promptly signs of gastrointestinal bleeding. Report blurred vision or any other visual abnormality (Velasquez and Wray, 2020).

Adolescents should not consume tablets whole before deferasirox is administered by nurse.. Completely disperse pills in water or juice (orange or apple). It is recommended that Adolescents take a tablet 30 minutes before eating and immediately after taking a suspension. If any residue is left, resuspend it in water and give it to your Adolescents to drink. Doses less than 1 g should be dispersed in 100 ml liquid; doses greater than 1 g should be dispersed in 200 ml liquid. Based on ferritin levels, treatment goals, and response, adjust dosage every 3 to 6 months in increments of 5 to 10 mg/kg every 3 to 6 months (Strachan and Greener, 2008).

If hypersensitivity reactions are extreme, the Adolescents should stop taking the medication and seek medical attention. When administering a diuretic, monitor the serum ferritin level monthly, the serum creatinine level, and the creatinine clearance weekly in Adolescents with additional renal risk factors. Consider dosage decrease, therapy interruption, or drug withdrawal if the levels rise (Piolatto et al., 2021).

The nurse teaching role is to advise Adolescents and family to avoid taking aluminum-containing antacids during therapy. Also, tell the Adolescents that the drug may cause vision and hearing disturbances, necessitating routine ophthalmic and auditory testing. If Adolescents has diarrhea or vomiting, notifying prescriber at time and staying hydrated (Ministry of Health and Social Welfare, 2013).

Nursing Diagnosis

- Knowledge deficient related to the cause of the Adolescents’s illness
- Anxiety related to frequent blood-sampling procedures

- Situational poor self-esteem due to the physical appearance change (pilliterrii, 2010).

2.5.1.j. Nursing interventions

The thalassemia patients like other patients with impaired and chronic illness, need prolonged lifelong care and deal with various mental issues and economic, social challenges, each engaging in every manner with the criteria and follow-up steps measures (Shahraki-Vahed et al., 2017).

The nurse performs a vital role in the patient's treatment with serious conditions like disorders of hemoglobin (Hb) such as Thalassemia and Sickle cell disease in every team of healthcare professionals involved. These illnesses are extremely challenging and if left untreated, they are fatal. These conditions can however be effectively managed by sharing knowledge expert about their treatment and prevention (Elewa et al., 2017).

Educate Adolescents and families regarding the prescribed routine. Ensuring families recognize that adherence to the recommended transfusion of the blood and chelation therapy regimen is necessary for survival of the Adolescents. A home regimen of chelation therapy is required to reduce iron levels in the body. Teach the members of family to administer deferoxamine subcutaneously over a period of several hours each night with a small, battery-powered infusion pump (usually while the Adolescents is asleep). If oral deferasirox is prescribed, advise the family to dissolve and administer the tablet once a day in juice or water (kyle & carman, 2013).

Both nurses and patients play an important part in empowerment. Patient education is an important aspect of nursing. It involves the enhancing patient's capacity to address problems, communication abilities and knowledge of health principles and empowers them to handle health problems. Self-management education program for chronic disorders has changed from the conventional framework of. The empowerment model, because patient education is successful at enhancing health performance, at least in the short term. Current counseling or approach to social work also lacks the non-medical (emotional, social, and cognitive) dimensions. Empowerment is a key to building effective communication with personnel- and patients. Empowerment is described as encouraging people to explore their innate capacity to manage their situation and disease (Razzazan et al., 2014).

To help the family and lessen the disease's symptoms, the primary goal of thalassemia nursing care is to provide for the Adolescents's physical and emotional needs. This entails administering blood transfusions and teaching the family. To keep the tissues supplied with oxygen and prevent erythrocytosis in the bone marrow, I administered packed red blood cell transfusions as instructed. Reactions to the transfusion should be closely monitored. Chelation therapy is used to remove excess iron (hemosiderosis) from the body. With the transfusion, administer Desferal (deferoxamine), a chelating agent. Iron can be excreted from the body through the urine or stool when Deferoxamine binds to the iron. It is possible to utilize Exjade (oral deferasirox), which is generally well-tolerated and has only minor gastrointestinal side effects (kyle & carman, 2013).

The specialists in nursing provide experienced, skilled support and encouragement throughout an often standardizes treatment regime, for this the pediatric nurse is an important element in the effective management of thalassemia. The nurse who is acquainted with the patients, families and their social situations is ideally positioned to provide an essential bind between the hematologist, the patient and other health professionals and necessary services. Nurses provide an acceptable standard of care (Elsayed & El-gawad, 2015).

The role of the nurse in encouraging adherence is stated to be to illustrate and explain both of thalassemia the complication of thalassemia and the consequences of non-adherence to chelation therapy. The most significant role of the nurse is to make the patient take an active part in his / her own care (Azize et al., 2015).

2.5.2.Sickle cell disease

Recurrent periods of vascular obstruction and inflammation characterize sickle cell disease, a genetic blood illness. SCD is a disease that affects multiple systems, resulting in both acute sickness and long-term organ damage (Rees et al., 2018). There are several genetic illnesses, but one of the most frequent is sickle cell disease. SCD affects over 200,000 infants every year, mostly in Sub-Saharan Africa (180,000 births per year), and it is responsible for about 3.5 percent of fatalities among Adolescents under the age of 5 (Cançado, 2012).

The blood illness sickle cell anemia (also known as sickle cell disease) is caused by an aberrant form of hemoglobin that runs in families (the oxygen-carrying protein within the red blood cells). Red blood cells become warped (sickened) as a result of the defective hemoglobin. The sickled red blood cells are prone to rupture because they are so delicate. Anemia occurs when the body's supply of red blood cells is depleted due to a rupture (hemolytic). Sickle cell anemia is the medical term for this illness.

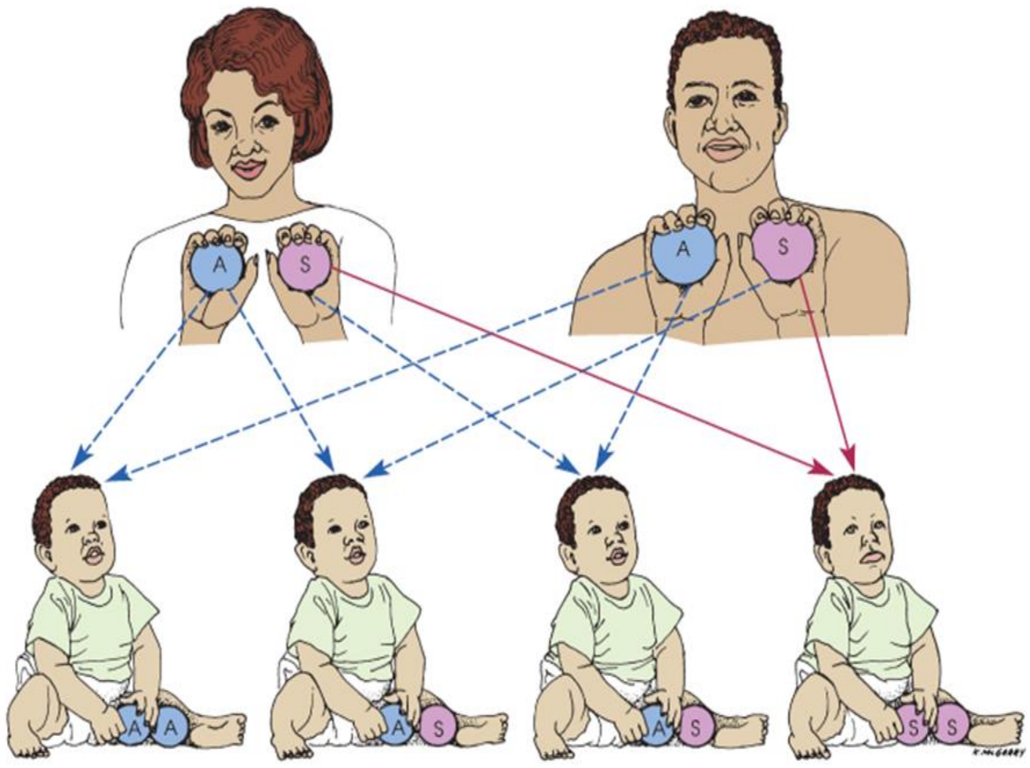
Blood vessels might get clogged as a result of the sucked cells, resulting in tissue and organ damage and pain (Ilesanmi, 2010).

2.5.2.1 Classifications of sickle cell disease

One of the most frequent forms of sickle cell disease (SCD) is sickle cell anemia (SCA), and the term SCD refers to all the numerous genotypes that generate the clinical condition. Co-inheritance of the S and C alleles causes Hemoglobin SC illness (Hgb SC). Hgb SBthal is the most common form of SCD, which arises when an individual inherits an allele of -thalassemia and an allele of S. (Al-Barazanchi et al., 2021).

A set of clinically relevant hemoglobinopathies known as sickle cell anemia is caused when the sickle gene is passed down from one parent to the next. There are two types of sickle cell anemia, homozygous and heterozygous. Sickle cell diseases can also be caused by the sickle gene being passed down in combination with the Hb C gene, resulting in Hb SC diseases, the sickle-thalassemia gene being passed down in combination with the sickle-thalassemia gene being passed down in combination with the sickle-thalassemia gene, and so on. These include Hb SS, which is the most common SCD form, as well as thalassemia, which is a compound heterozygous disorder (El-Hazmi et al., 2011). It is usual to refer to Hb SS and HbS0-thalassemia as SS or SCA because these genotypes are linked with the most severe clinical symptoms. Certain SCD problems may be more prevalent in one category than in another. As a result, SCA sufferers are more likely to experience many painful episodes, severe anemia that necessitates blood transfusion, and acute chest syndrome (ACS). When it comes to eye and ear problems, Hb SC illness is more common than SCA.

The severity of SCD is depending on demographic statistics, so it's important to emphasize this. To put it another way, when comparing 100 people with SCA and Hb SC disease for example, the latter will have a lower frequency of unpleasant episodes, morbidity, and mortality than the former. There are, however, exceptions to the rule when it comes to unique cases. A patient with SCA may have a minor condition, while an S-+-thalassemia patient may have a more severe one (da Guarda et al., 2020).



2.5.2.2 Sickle cell disease pathophysiology

Hb S crystallizes and forms polymers of haemoglobin in deoxygenated situations. The crescent-shaped RBCs are distorted as a result of this. Small blood arteries are blocked by sickled RBCs, resulting in painful vaso-occlusive crises (Li et al., 2017).

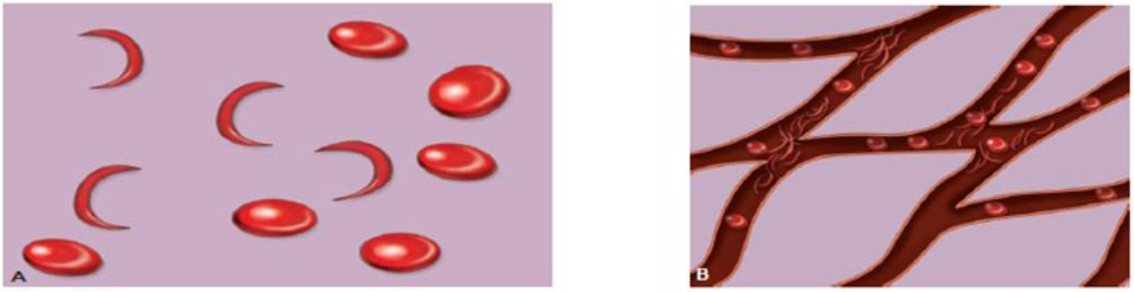
Sickle cell illness can be passed down via a family. Hemoglobin A (the normal type of hemoglobin) in the parent's red blood cells protects them from hemoglobin S, which causes sickle cell disease in their Adolescents (the sickling form). It's possible, however, for two carriers to become parents. Sickle cell disease (SS) affects one in every four Adolescents who inherits all of their parents' hemoglobin, but only one in every two Adolescents inherits both their parents' hemoglobin A and their parents' hemoglobin S, making them both carriers of the trait (Leifer and Keenan-Lindsay, 2019).

Simplified genetic scheme for sickle cell disease. A denotes adult hemoglobin; S denotes sickle hemoglobin. Hemoglobin AA = normal hemoglobin, hemoglobin AS = sickle cell trait, hemoglobin SS = sickle cell disease (Kyle, 2008).

HBS is caused by a substitution of thymine for adenine in the sixth cordon of the beta chain gene, GAG to GTG. This results in the coding of valine rather than glutamate in the HB beta chain at position 6. Deoxygenation gives rise to HB with polymerization capabilities. The solubility and molecular stability of the substance are also altered.. It is because of these characteristics that sickling syndromes exhibit such severe clinical manifestations (Olubiyi et al., 2019).

Sickle-shaped red blood cells and impaired hemoglobin function characterize hemoglobin SS illness in Adolescents. The progression of sickle cell disease is influenced by factors such as elevated body temperature, high hemoglobin concentration, low blood pH, and dehydration. The sickling of erythrocytes might be reversible or permanent. Because of their sickle-like structure, the first group loses oxygen, while the second group retains it (Bowden and Greenberg, 2010).

Shockingly, sickle cells have a rigidity that makes it difficult for them to adapt to their environment, particularly in the microvasculature. (Fig.). Premature hemolysis of sickle cells results in chronic anemia. In addition, sickle cells aid in the enhanced adhesion of sickle cells to the vascular endothelium that results from vaso-occlusion in sickle cell disease (Connes et al., 2016).



In sickle cells, faulty hemoglobin causes the cells to exhibit a sickle-shaped appearance. As a result, blood arteries get clogged with hardened cells. Pain, infection, and organ damage can result from blocked blood arteries (vaso-occlusive crisis).

2.5.2.3Clinical manifestation of sickle cell disease

More than 90% of all SCD-related hospitalizations are due to pain, which is the disease's defining symptom. Infection, dehydration, changes in climate, and physical and emotional stress can all lead to a vaso-occlusive crisis. Asthma, acute chest syndrome (ACS), renal disease, and cerebrovascular disease (CVD) are only a few of the medical conditions that SCD patients are at risk for (Borhade, 2019).

The clinical manifestations of SCA can vary widely in severity and frequency. During the first 6 months of life most newborns are asymptomatic because Hb F does not sickle. As Hb S replaces Hb F, however, sickling increases and symptoms begin to develop. The anemia of SCA is manifested by the same signs and symptoms of other anemias, including pallor, fatigue, shortness of breath, irritability, and jaundice. Adolescents with SCA also experience delayed growth and delayed onset of puberty (Akinsheye et al., 2011). However, the principal symptom experienced by Adolescents with this disorder is pain. This pain can occur unpredictably in any body organ or joint and is typically associated with the occurrence of vaso-occlusive crises. These crises are the most common type of sickle cell crisis, although sequestration crisis and aplastic crisis can also occur (Ballas et al., 2012).

Because sickling is hindered by high amounts of fetal hemoglobin, affected Adolescents are normally asymptomatic until they are 4 to 6 months old. Clinical manifestations are directly related to the shortened life span of blood cells (hemolytic anemia) and tissue destruction resulting from vaso-occlusion (blockage of a blood vessel). Illness results from recurrent vaso-occlusive events that involve painful crises and chronic organ damage. Pathologic changes happen in most body systems, resulting in multiple signs and symptoms (Telen et al., 2019) . Examples of common organs affected include the following:

- Brain—stroke, often manifested by headache, aphasia, convulsions, visual changes
- Eyes—retinopathy, retinal detachment, diminished vision
- Bones—chronic ischemia of bones with susceptibility to infection and bone degeneration, manifested by osteoporosis, osteomyelitis, spinal deformities, or aseptic necrosis of the femoral head
- Liver—impaired blood flow from capillary obstruction leads to enlargement and scarring of the liver, manifested by hepatomegaly or cirrhosis
- Spleen—splenic infarct leads to fibrosis and increased rates of infection
- Adolescentsneys—ischemia of Adolescentsneys causes enuresis, hematuria, inability to concentrate urine
- Penis—microcirculatory obstruction and engorgement (priapism)
- Extremities—vaso-occlusion and chronic ischemia manifests as peripheral neuropathy, weakness, or arthralgia

Skin—decreased peripheral circulation causes ulcerations(Ball et al., 2017).

2.5.2.4 Management of sickle cell disease

2.5.2.4.a Vaso-Occlusive Pain

Effective SCD management requires early access to a comprehensive range of preventive screenings, urgent care treatments for vaso-occlusive pain crisis and ongoing prophylactic treatments such as blood transfusions or hydroxyurea. More than 200,000 patients with SCD are seen in the emergency department each year in America for vaso-occlusive crises. The study results revealed that the severe pain scale used during triage showed 54% of patients with SCD rated their pain between 7 and 10 on an 11-point scale (Kanter and Kruse-Jarres, 2017).

Adolescent with SCD require frequent hospital care for painful vaso-occlussive crises. Elevated rates of inpatient hospitalizations for sickle cell crises are associated with poor outcomes and low quality of life for both patients and families. So, the hemoglobinopathy nurse have a vital role in pain management for Adolescents with SCD . The use of patient-controlled analgesia (PCA) dosing is determined based on the influence of chronic anemia opioid pharmacokinetics in SCD (Wilkie et al., 2010).

2.5.2.4.b Blood Transfusions

Vaso-occlusion problems in SCD are alleviated by red blood cell transfusions. Both acute and long-term therapy of SCD relies heavily on blood transfusions. Transfusion can be life-saving in some acute conditions (e.g. ACS, multi-organ failure, acute anemic episodes) Although there have been few controlled trials to guide acute transfusion therapy, several major research have served to define the significance of chronic transfusion, notably in the prevention of primary and secondary cerebral ischemia injury (Howard, 2016).

Primary and secondary prevention of sickle cell disease are two separate but related goals of transfusion. In SCD, blood transfusions can be dangerous, and some consequences are difficult to avoid. In each scenario, the risk/benefit ratio must be carefully evaluated. Infection, all immunization, and post-transfusion hyper hemolysis are the main dangers associated with transfusion in SCD (Davis et al., 2017).

2.5.2.4.c Hydroxyurea

Long-term control of SCD is currently only possible with the approved medication Hydroxyurea. In 1998, the FDA approved the use of hydroxyurea for the treatment of people with clinically severe SCD. There is a strong recommendation regarding use hydroxyurea for infants and Adolescents 9–42 months. The Initial dose as 15 mg/kg once a day that may increase by 5 mg/kg/day every 12 weeks till the maximum dose of 35 mg/kg/day (Strouse and Heeney, 2012).

During crisis situations, -treatment is largely supportive and symptomatic. Oxygen administration and the use of analgesics and sedatives seem to be somewhat effective in reducing the painful symptoms of the acute sickling periods. Management and care must extend beyond medical treatment. Optimal nutrition, satisfactory dental and hyenic care, and avoidance of exposure to infection an~ crisis precipitating factors are essential in the care of the individual during quiescent periods (Adewoyin, 2015).

Supportive and symptomatic care is provided for infants and Adolescents who are hospitalized during a crisis. Restraints are placed on the patient. To alleviate discomfort, analgesics are offered. A narcotic intravenous (IV) infusion may be necessary for Adolescents who are in excruciating pain.

There is a risk of nor meperidine-induced seizures with meperidine (Demerol) in Adolescents with sickle cell illness. Control and participation in care can be maintained with the use of a patient-controlled analgesia (PCA) pump for Adolescents over the age of 7. To prevent dehydration and acidosis, all efforts are made. Hemoglobin levels can be temporarily raised by small blood transfusions, although this is a short-term solution. For this reason, pre-packaged RBCs are frequently distributed. The intake and output of food and water are meticulously tracked. The position of the patient's body is constantly yet softly modified. Soon, a treatment may be found through the use of blood stem cell transplantation (Leifer, 2013).

A sickle cell crisis can result from a number of factors, including dehydration, hypoxia, and infection. Fetal hemoglobin synthesis can be increased by erythropoietin and various chemotherapy regimens. Patients with leukemia may benefit from bone marrow transplantation and gene therapy (Ashorobi and Bhatt, 2019).

2.5.2.5Consequences of sickle cell anemia

Pupil and adolescent years are connected with several acute and chronic consequences of SCD, such as the sequestration of an individual''s spleen, renal impairment, unpleasant sexual experiences, avascular necrosis, and an increase in vaso-occlusive episodes. Sickle-related structural and physiological abnormalities can lead to Adolescentsney problems. One in ten school-aged Adolescents with SCD will experience a cerebral vascular event (a stroke or a silent infarct) (Tanabe et al., 2019). Vaso-occlusive events are more common in adolescents who engage in more physical activity. Adolescent men who are either homozygous for Hb S (Hb SS) or heterozygous for Hb S and -thalassemia are the most likely to develop priapism, with an incidence of 89 percent. SCD patients experience a two-year average delay in puberty because to their slower growth than youngsters with a normal Hgb genotype (Mantadakis et al., 2009).

2.5.2.6 Prevention of sickle cell disease

Prevent and control of SCD is still lacking in most countries of high prevalence (WHO, 2008). Current research supports the idea that SCD is preventable if individuals are aware of their sickle cell trait status and undergo genetic counseling (Pecker and Naik, 2018).

Adherence to premarital screening can lead to decrease in getting an affected Adolescents with long lasting debilitating illness. It eliminates health and cost of frequenting the hospital for treatment. Sickle cell disease cases will go down, as will the high newborn mortality and morbidity rates that these Adolescents endure (Anunobi, 2014).

2.5.2.7 Nursing management of sickle cell disease

Nurses play a pivotal role in delivering day-to-day and emergency care and good feedback will help nurses understand the impact of the care they give and be lauded where they are doing well, while allowing improvements if they are necessary (Shalala et al., 2011).

The best-practice guidelines for nurses should include the following: rapid clinical assessment of vaso-occlusive crises, use of an age-appropriate pain scale, pharmacologic management as prescribed by a medical doctor; monitoring Adolescents's pain every 20 minutes until pain is controlled and stable; and monitoring vital signs, hydration, and oxygen saturations (Dunlop and Bennett, 2009).

Family members should be informed of the prescribed regimen for their Adolescents. Families need to be made aware of the need of sticking to the prescribed blood transfusion and chelation therapy regimen. In order to keep the iron levels in the body at a safe level, chelation therapy must be continued at home. Teach family members how to inject deferoxamine subcutaneously for several hours each night using a small battery-powered infusion pump (usually while the Adolescents is sleeping). If you've been prescribed oral deferasirox, make sure to explain to your loved ones how to take it and how often (Verissimo et al., 2013).

2.5.2.7.a Family education and Discharge Instructions for Adolescents with SCA

When a adolescent or family member is diagnosed with SCD, they and their loved ones must learn how to live with the illness for the rest of their lives. When a sickle cell crisis occurs, families must be aware of the warning signs and symptoms so they can notify their primary care practitioner. It is possible to communicate with the patient's family about long-term care goals, such as preventing infections, hypoxemic episodes, and a vaso-occlusive crisis. It's important to make parents aware that vigorous activities can lead to dehydration and hypoxia, therefore they should avoid them at all costs. As a precaution, parents should be aware of probable problems, such as pain and fever management, splenic sequestration, priapism, acute chest syndrome, and even a stroke. Almost any of these problems could lead to long-term impairment or death (Ward and Hisley, 2015).

Sickle cell crisis can be avoided by teaching parents how to provide appropriate rest and hydration. Prophylactic penicillin is given to SCD patients to help prevent sepsis and supplementary folic acid is given to help with red blood cell synthesis in these patients. Prevention of illness consequences requires strict adherence to these drugs. In addition to scheduled medications, all required immunizations should be administered per AAP guidelines. Parents must be aware that preventable illnesses have the potential to be life threatening for a patient who is a splenic (Yawn and John-Sowah, 2015).

A moderate sickle cell crisis can occur at home; urge the patient to stop what they're doing, drink fluids, and take the recommended pain medicine if this happens to him or her. If the patient's condition doesn't improve after a few days, call their doctor or take them to the nearest hospital emergency room for further treatment (Esham et al., 2020).

2.5.2.7.b nursing care plan for Adolescents with SCA.

Nursing diagnosis:

1-Ineffective Tissue Perfusion related to vaso-occlusion and anemia

Administer oxygen as prescribed. Monitor oxygen saturation, capillary refill, and respiratory status. Maintain the client in a position of comfort (semi- to high-Fowler’s). Administer packed RBCs as prescribed (Luxner and Comer, 2009).

2- Acute Pain related to vaso-occlusion and tissue ischemia

Apply heat to affected area; avoid cold compresses. Administer analgesics as prescribed (preferably morphine) and assess effectiveness by assessing behavior and vital signs. Reassure the caregivers that opioids are appropriate, high doses may be needed, and addiction is rare. Position the client carefully and handle gently (White et al., 2019).

2.6. Quality of Life(QOL).

2.6.1. Quality of Life Overview

The concept of Quality of Life lacks a clear definition. It has been used within medical researches to refer both to the measurable, material conditions of life and to the experiences that make life meaningful and valuable (Post, 2014).

A good quality of life is a state of physical and psychosocial wellbeing, in which the individual is able to perform everyday activities and reports satisfaction with daily function. There are several studies reporting a significantly lower quality of life in thalassemia patients compared to healthy controls (Khodashenas et al., 2021).

The term Quality of Life refers to the physical, psychological, and social domains of health, seen as distinct areas that are influenced by the person’s experiences, beliefs, expectations, and perceptions. The quality of life is not a new concept, many sciences such as sociology, psychology, and economics have used it. The term quality implies the degree of excellence of characteristics, different people may value different areas of life, and therefore Quality of Life means different thing to different people (Theofilou, 2013).

Also the quality of life , can been seen differently by individuals who may be affected with economic hardship, declining health, and social losses. Individuals who have good health, have money and a high social status can perceive life to be empty, lonely, and without quality. Objective quality of life refers to Health-Related Quality of Life (HRQoL) which is the domains that reflect health and health care effects. Subjective Quality of Life also focuses on health-related Quality of Life research. Subjective quality of life defined as the fit between a person’s expectations and his or her achievements (Singh and Singh, 2008).

The WHO (1998), considers that the Quality of Life is an individual’s perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals expectations, standers and concerns, there are a variety of terms equated as QoL, Life satisfaction, wellbeing, health, happiness, and adjustment, value of life, functional status and meaning of life (WHO, 2012).

A moderate sickle cell crisis can occur at home; urge the patient to stop what they're doing, drink fluids, and take the recommended pain medicine if this happens to him or her. If the patient's condition doesn't improve after a few days, call their doctor or take them to the nearest hospital emergency room for further treatment (Esham et al., 2020).

In clinical trials, quality of life measures can be used to assess the relative impact of different diseases, to gauge treatment effectiveness, to gauge the standard of care, and to pinpoint individual patients' specific problems and gauge the effectiveness of treatment to address those issues. Quality of life measures can also be used to assess the general quality of life of populations (Ware et al., 2016).

There are two types of quality of life measures: generic and disease-specific. Measures of general health-related quality of life are sometimes referred to as "generic." It is essential that their content be wide-ranging and encompass those areas of Quality of Life that are most significant to the majority of people, such as physical and social function and pain relief (Hand, 2016). 2.10.2. 2.14.0.

2.6.2 Quality of Life domains

Varni, et al.,2004 the Pediatric Quality of Life Inventory measurement model is a modular approach to measure health-related Quality of Life (HRQoL) in healthy Adolescents and adolescents and those with acute and chronic health conditions PedsQL version 4.0 consists of Physical, Emotional, Social and school functioning.

2.6.2.1Physical domain

Thalassemia has a negative impact on Quality of Life due to the effects of the disease and its treatment, not only affecting Adolescents's physical function, but also their social, emotional, and school function, leading to an impaired Quality of Life (Thavorncharoensap et al., 2010).

Thalassemia is a long-term condition that comes with a slew of medical and mental difficulties. Physical deformities, growth retardation, and a delayed onset of puberty are all possible outcomes of thalassemia-related health issues. Physical appearance, such as bone abnormalities and small stature, contributes to low self-esteem. Many thalassemia patients suffer from serious side effects, including heart failure, arrhythmias, liver disease, endocrine problems, and infections. Patients' physical, emotional, social, and educational functioning are all affected by the issues described, which results in a decreased HRQoL (Health-Related Quality of Life).

Thalassemia major presents itself clinically between six and 24 months of age. Infants that have the condition are underweight and pallid as they grow older. When the spleen and liver grow, so does the size of the belly. This can lead to issues with feeding, diarrhea, and irritability (Ikehara, 2011). Thalassemia major is characterized by growth retardation, pallor and jaundice, poor muscle tone, genu valgum, hepatosplenomegaly and leg ulcers, the development of masses from extramedullary hematopoiesis, and skeletal changes due to the expansion of the bone marrow in some developing countries where resources are limited. Changes in skeleton and craniofacial features include bossing of the skull, a pronounced malar eminence, a slanted eye, and hypertrophy of the maxillae, which tends to expose the upper teeth, among other things (Origa, 2017).

2.6.2.2 Emotional functioning

Emotional QOL was affected because thalassemia Adolescents feel different from their peers and elaborate negative thoughts about their life. Adolescents at this age are becoming more aware of themselves as individuals. They work hard at being responsible, being good and doing it right (Eldakhakhny, 2011).

They are now more reasonable to share and cooperate but the disease prevents them from being industry and the sense of inferiority may develop instead . Thalassemia major and its repercussions have a particularly negative effect on adolescents' mental health, resulting in feelings of pessimism and isolation as well as trouble adjusting to social situations (Shamsi et al., 2017).

2.6.2.3 Social functioning

Adolescents affected by thalassemia are unable to participate in outdoor play at the same level as their classmates due to the disease's physical weakness. Sporting activity were therefore restricted. Sixty-eight percent of the young people surveyed said they were unhappy with the way they looked. Almost majority of them stated their dissatisfaction with their physical appearances as the primary factor. Self-esteem was negatively affected due to a lack of growth, physical anomalies, and a delayed onset of puberty.

Chronic disorders, such as thalassemia, can lead to feelings of inadequacy and self-doubt, as well as an increased need for assistance. The growth of bones, notably the skull and jawbones, is responsible for the thalassemia-related facial features. Short stature and delayed puberty are common side effects of anemia and iron excess in these people. Five thalassemia teenagers were interviewed in open-ended fashion about the disease's effect on their sense of self-worth. The majority of them were afraid of being rejected or mocked for their appearance. There were 45 adolescents in our study (91%), all of whom were concerned about their physical appearances (Pinto and Forni, 2020).

2.6.2.4 School functioning

The school functioning subscale scored the lowest. This could be explained by the fact that frequent absenteeism from school for hospital visits, and a lack of energy when performing academic activities, had a significant negative impact on the patient Health Related Quality of Life HRQoL (Ismail et al., 2009).

The validity of PedsQLTM SCD has been proven by numerous studies conducted in various locations throughout the globe. SCD-specific health-related quality of life can be assessed in clinical research and practice using the PedsQLTM SCD Module, especially when paired with the PedsQLTM Multidimensional Fatigue Scale. When it comes to the quality of life of school-aged Adolescents 6 to 12 years of age with thalassemia major, an Iranian experimental study was conducted (Panepinto et al., 2012). Surveys on Adolescents's demographics and general well-being, including information on their physical, emotional, and social well-being as well as academic performance, were used to compile the data. Following confirming the validity and reliability of the questionnaires using the internal correlation approach, quality of life was evaluated 1.5 months after the intervention. After the intervention, the quality of life of thalassemia youngsters was greatly improved. In spite of this, the questionnaire wasn't used in any subsequent studies to test its efficacy (Al-Blaihed et al., 2020). According on psychometric measurements, Greek researchers looked at the needs of adult patients with thalassemia and developed a self-administered STQOLI. First, a qualitative study was carried out to generate items and identify domains using the critical analysis incident technique and a literature review, this qualitative study involved both patients and experts, then quantitative validation was carried out to select items, identify dimensions, and measure reliability and internal and concurrent validity, finally the questionnaire had 41 items comprising four main domains and one global item about general hippocampus functioning. The HRQOL of patients with thalassemia can be assessed using this instrument, however further research is needed to further grasp the questionnaire's universal features (Mahmoud, 2015).

The HRQOL and the Pediatric Quality of Life Inventory assessment for thalassemia Adolescents in Jordan showed the lowest mean score in the school and physical functioning areas. To corroborate the findings, a comparison was made to healthy youngsters with similar demographics and clinical data (Azarkeivan et al., 2009).

The PedsQL4.0 was selected for this investigation because it has been shown to be acceptable reliability when judged by internal consistency and is valid and reliable for use with chronically ill Adolescents and healthy Adolescents ages 2-18. When used in a clinical environment, the PedsQL4.0 is simpler to administer since it is all of the above: short, easy-to-use, adaptable, developmentally appropriate, multidimensional, and all of the above. The WHO's primary health dimensions were taken into consideration when developing the PedsQL4.0 generic core scales. Physical, emotional, educational, and social functioning are all included in the PedsQL4.0. While the CHQ caregiver report form analyzes the familial and parental impact of a Adolescents's health, the PedsQL4.0 contains a social functioning subscale that is used to assess peer connections (Wrotniak et al., 2014).

The PedsQL4.0 has been found to be a reliable tool for measuring Adolescents's quality of life. The PedsQL4.0 also has a caregiver report version for young Adolescents and two questionnaire lengths: long and short. Comparisons between caregivers and Adolescents will be easier with separate tools for each group. The PedsQL4.0 has a restriction in that Adolescents and caregivers are asked to recollect information from the previous month. For example, if the Adolescents has not been to school recently, the Adolescents and caregiver may not be able to answer school-related questions (Desai et al., 2014).

A last consideration was that Adolescents can report on their QOL in a clinical context, which provides information about their daily functioning that may have an impact on QOL. It was chosen for this study because the PedsQL4.0 may measure quality of life in Adolescents and adolescents with acute and chronic health problems, respectively. The PedsQL4.0 questionnaire's generic and disease-specific scores can be merged into a single measurement system (Wu et al., 2013).

2.6.3 Importance of quality of life

Quality of life can be used to define the health profile and functional state of Adolescents with the disease in order to identify the impact of sickness. 4 In the evaluation of new medical techniques, such as transfusion therapy and bone marrow transplant, the information presented can be useful. Adolescents with SCD who are taking new drugs (hydroxyurea) may benefit from this information. Adolescents with SCD and their families' quality of life can be assessed as well. These findings would lead to the creation of appropriate therapies to target areas of functioning where there is a deterioration in quality of life (Bhatia, 2011).

2.7. Behaviors:

It is common for people with SCD and their families to suffer from psychosocial issues as a result of the everyday impact of pain and other SCD symptoms on their life. Psychosocial issues might arise from people's perceptions of SCD and their treatment of patients with the condition. In comparison to their healthy peers, Adolescents and adolescents with SCD are more likely to experience poor academic outcomes due to missed school days and hospitalizations. In addition to physical pain, SCD pain crises can cause mental health issues such as sadness and anxiety. As a result of these mental problems, people are less able to deal with discomfort (Anie et al., 2010).

More than a quarter of Adolescents with SCD also suffer from symptoms of depression and anxiety, as well as difficulties with social interaction and low academic achievement. Adolescents with SCD have an increase in the challenges of daily life. These feelings of pessimism, despair, and social isolation can occur in teenagers with SCD who are more easily fatigued when participating in sports. Many Adolescents with SCD have low self-esteem and depressed symptoms, particularly because they are unhappy with their body's look, due to a lack of growth and delayed sexual development. Adults who have SCD may suffer from a variety of psychological side effects, including concern about one's appearance, treatment, and death, as well as sadness, guilt, and low self-esteem. Anxiety, sadness, and suicide ideation are all signs of depression in people with SCD (Pandarakutty et al., 2020).

The influence of family, friends, and school can all have an impact on a Adolescents's capacity to engage in healthy or dangerous behaviors, and in some cases, SCD itself can have a negative impact on a Adolescents's ability to engage in healthy behaviors or encourage them to engage in risky behaviors (Trost and Loprinzi, 2011).

Emotional anguish is a risk factor for more involvement in these behaviors, and perhaps their short lifespans indicate that they are more impulsive and want to experience life to the full. Alcohol, smoking, and illegal drug use were all shown to be common risky behaviors among teenagers with chronic illnesses, according to a recent analysis. Adolescents' self-report measures of risky behavior are common, although these may not always be reliable and under-reporting happens. As a result, parents' reports may not be effective because they are unlikely to be aware of their Adolescents's harmful behaviors and objective assessments are difficult to collect (Balocchini and Chiamenti, 2013).

Psychologically, -thalassemia major and its sequelae can be particularly devastating in youngsters, causing them to feel helpless and socially isolated. According to the literature, thalassemia youngsters have reduced abstract reasoning, deficiencies in language, attention and memory, constructional/visual spatial skills, and executive functions. When it comes to the severity and chronicity of the condition, a low intelligence quotient appears to be linked to poor school performance and physical or social limits for the severity and duration of the illness (Mazzone et al., 2009).

The behavioral profile of thalassemia adolescent is similar to that of normal subjects, but many of them can manifest severe psychosocial problems due to difficulties in complying with the painful chelation; male patients, in particular, show oppositional defiant disorder. Thalassemia Adolescents feel different from their peers and elaborate negative thoughts about their life, guilt senses, increased anxiety and low self-esteem. Relatives may become more distant and isolated as a result of their fears for the future of the Adolescents's thalassemia condition (Saini et al., 2009).

Adolescent with thalassemia are at risk for emotional and behavioral issues because of their long-term medical conditions. Anxiety and depression can put an enormous strain on a Adolescents's emotional and interpersonal resources, as well as on their families. Thalassemia patients' emotional and cognitive requirements are clearly distinct from those of adolescents who are attempting to establish their own identities. Thalassemia patients experience difficulties in finding a partner, starting a family, and losing social support as their parents get older (Shaligram et al., 2008). As a result, difficulties specific to each age group arise. However, as the lifespan of Adolescents with thalassemia has increased, so has the prevalence of mental health issues. As many as 80% of thalassemia Adolescents’ experience mental health issues, including obstructive teen disorder, anxiety disorders, and depression. Adolescents with thalassemia must go to the hospital for blood transfusions on a regular basis, which makes them unusual among Adolescents with blood disorders. According to research on thalassemia quality of life in teenagers, individuals with mental health issues had a lower QOL (Behdani et al., 2015).

During the time period from January 1, 2009, to March 1, 2018, a large number of empirical studies revealed that parents' experiences included physical and psychological distress as well as social isolation and social alienation, as well as sleep problems, weakness, fatigue, and a failure to take care of one's own needs. 6-10 Sixty-six percent of careers for adolescent with thalassemia were found to have little satisfaction in their lives, and eighty-two percent expressed fears about the future for the Adolescents.9 (Punaglom et al., 2019).

Adolescent are at high risk of developing behavioral and psychosocial problems like opposition, passiveness, anxiety, phobias and depression, which affect their self-confidence and give rise to emotions and thoughts which negatively affect their quality of life and compliance to therapy (Beesdo et al., 2009). Many of the thalassemia Adolescents experience fear related to intravenous line insertion and subcutaneous infusion pumps. Adolescent with thalassemia in the pre-school and latency age groups are usually anxious and excessively dependent on their parents. They display psychosomatic symptoms and are frequently absent from school. Thalassemia Adolescents have more of negative self-concept when compared to their normal counterparts (Kareem, 2014).

Adolescent with SCD face a slew of challenges in the classroom. Stroke-related memory loss, a decline in academic performance in reading, math, spelling, and science, are just a few of the challenges students face. SCD patients frequently miss school owing to frequent hospitalizations (Strunk et al., 2020).

A total of 24 adolescent with SCD were studied for their body satisfaction, sadness, and social withdrawal. Adolescents with SCD were shown to be less satisfied with their physical appearance. They also discovered that these adolescent had greater symptoms of depression and spent less time participating in both social and non-social activities than their healthy peers. These difficulties could have a significant impact on their academic success. As a result, effective interventions are required for these students (Bhatt-Poulose and colleagues, 2016).

Adolescent who suffer from long-term illnesses are more likely to experience behavioral or emotional issues. While caring for a sick Adolescents can be emotionally and physically taxing for both parents, it can also put a strain on the family's finances (Ogundele, 2018).

The nurse works with the family to determine whether or not the Adolescents's daily routine needs to be normalized. Adolescents who spend most of their time in their bedrooms need a reconstructed daily routine to provide them access to other parts of the house, such as the kitchen or dining room. A normalization approach can let these Adolescents participate in a variety of activities that they may otherwise miss out on. As an illustration, health-related therapies administered at home or away from home should be scheduled at times when they will have the least impact on everyday activities (Hockenberry and Wilson, 2013).

There should be a greater emphasis on the positive aspects of a person's appearance and abilities for Adolescents who feel self-conscious about having a disability. In order to build a positive self-image, health experts encourage youngsters to express their feelings of anger, isolation, fear of rejection and loneliness while highlighting the normal. Compliance and any signs of improvement among the Adolescents necessitate positive reinforcement. In order to improve one's self-esteem and self-confidence, people will do whatever to enhance their appearance and boost their self-esteem, whether it is through the use of make-up, clothing to hide prosthetics, or a new hairdo or wig (Perry et al., 2017).

Adolescence can be a particularly difficult period for the teenager and family. All of the needs discussed previously apply to this age group as well. Developing independence or autonomy, however, is a major task for the adolescent as planning for the future becomes a prominent concern. Although the emphasis in the past has been on achieving independence from physical assistance, recent developments in the fields of special education, adolescent development, and family systems suggest redefining autonomy in terms of individuals' capacities to take responsibility for their own behavior, to make decisions regarding their own lives, and to maintain supportive social relationships (Hockenberry and Wilson, 2018). Given this understanding, even individuals with severe impairments can be viewed as autonomous if they perceive their own needs and take responsibility for meeting them, either directly or by engaging the assistance of others. As adolescents become more autonomous, the nurse can help them articulate their needs, participate in developing their own care plans, and discover and express how others can be of greatest assistance (Speedie and Middleton, 2021).

Acquiring one's identity and developing one's sense of self-determination occur during adolescence. Knowledge, abstract thinking, and reasoning enhance during this transitional period of physical and psychological growth as a result of puberty. Peer groups are formed, parents are separated (emotionally and physically), one's physical appearance is established (e.g., clothes, haircuts), one's interests and hobbies are explored, one's self-confidence is developed, and one's identity is established (Erwin, 2021). Individuals may also begin to explore their sexuality or pursue romantic relationships during this time period. Adolescent social isolation or loneliness might seem as a lack of social connections or communication with others. During adolescence, an individual's social life undergoes a dramatic shift. More time is spent socializing with friends rather than family members during this period. Teenagers' perceptions of what it means to be alone are changing in response to this transformation. In fact, spending time with parents or other relatives may be seen as social isolation by others. Feelings of loneliness can be exacerbated in teenagers who believe that being isolated from their peers is a sign that they are unpopular (Kar et al., 2015).

Physical symptoms are high on teenagers' list of health-related concerns. Because adolescence is a time of enormous physical and emotional changes, it is important for the nurse to distinguish between body changes that are related to the Adolescents's complex condition and those that are a result of normal body development. It can be a great comfort for teenagers with disabling conditions to know that many of the changes they experience are normal developmental outcomes. A sense of feeling different from peers can lead to loneliness, isolation, and depression. Participation in groups of teenagers with chronic conditions or disabilities can alleviate feelings of isolation and smooth the transition to a meaningful relationship with one person in adulthood (National Academies of Sciences, Engineering, and Medicine, 2019).

2.8 Theoretical framework:

The RAM's theoretical underpinnings. Sr. Callista is As a PhD student at the University of California, Los Angeles, Roy began working on the RAM in the mid-1960s. An experimental design for testing one aspect of the concept of nursing as "promoting patient adaptation" was proposed by Roy in her term paper entitled, A Design for Testing One Aspect of the Concept of Nursing as the Promotion of Patient Adaptation. Roy's idea of adaptability was built on this foundation (McEwen and Wills, 2017). As early as Roy's (1965) model work, it was noted that a nurse's role is to assist and promote adaptability in patients who are ill or at risk of becoming sick. The importance of nursing in appraising a patient's ability to cope with disease was also brought to light (Roy, 1965). A nurse's distinctive role in providing patient care and addressing society's health-care demands was first outlined by Roy in the early 1970s (Russo, 2019).

The theoretical framework for QOL from the selected concepts of Roy adaptation model was formulated in Figure 1. The theoretical propositions were formulated based on the RAM and relevant theoretical and empirical findings from the literature. In particular, RAM propositions were tested in this study.

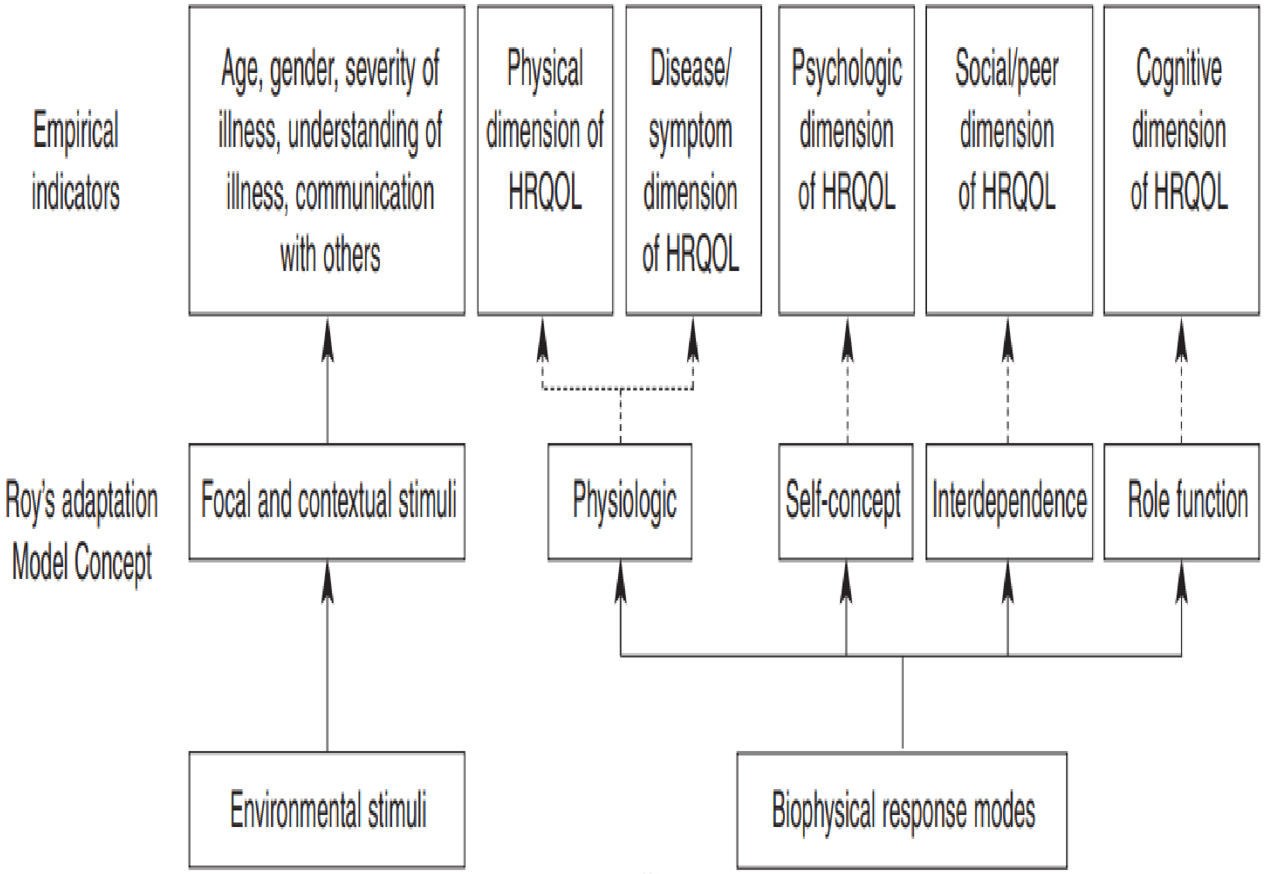


Figure 1 ■ Conceptual structure for health-related quality of life.

Modified from (Naga and Al-Atiyyat , 2014).

1. The environmental stimuli (severity of illness, age, gender, understanding of illness, and communication with others) influence the biopsychosocial responses of cancer Adolescents.

2. The 4 response modes from RAM (physiologic, self-concept, interdependence, and role function), that is, 5 dimensions from HRQOL (physical, disease/symptom related, psychologic, peer/school, and cognitive) are interrelated.

2.9. Previous studies:

1- Researchers (AlJabiri, et al., 2015) conducted a study called (Assessment of Quality of Life for Patient with Thalassemia at Thalassemia Center in Thi-Qar Province). Objective: To Assessment Quality of Life for Patient with Thalassemia at Thalassemia Center in Thi-Qar Governorate, and to identify the association between nurses' socio-demographic characteristics of age, gender, marital status, occupation and blood group and their effect to quality of life for patient with thalassemia.

Methodology: A purposive "non-probability" sample of (100) patient who were selected from thalassemia Center in Thi-qar governorate for the period from October 4th 2014 to April 1st of 2015. The study results indicated that more than half of the sample participants are resident urban areas (58%), more than half of the sample participants were men (58%), more than half of the sample of participants are from the age group (5-10) years (54%), that the vast majority of the participants were single (91%), one-third of the sample almost with low literacy (38%), most of the sample jobless (78%) and there is a significant relationship between the quality of life and some of the variables (such as age, level education and occupation).

2- Health-related Quality of Life in Adolescents and Adolescents With b-Thalassemia Major on Different Iron Chelators in Basra, Iraq was conducted by (Abdul-Zahra et al., 2016). A case-control study was performed on patients with b-thalassemia major registered at the Center for Hereditary Blood Diseases in Basra from February 2012 through July 2013. The group included Adolescents and adolescents aged 2 to 17 years old. HRQoL was assessed using the Pediatric Quality of Life (PedsQL) Generic Core Scale questionnaire, version 4.0, for Adolescents 2 to 12 years old and the Short Form-36 health survey questionnaire, version 2 (SF-36v2), for Adolescents and adolescents aged 13 to 17 years old. A total of 209 age-matched and sex-matched Adolescents and adolescents were included in the control group. The study did not find a significant difference in PedsQL scores among different age groups or different iron chelators, whereas there were significant differences in all of the SF-36v2 domains, with the best quality of life observed in the deferasirox group, followed by the deferoxamine group and the combined therapy group ($P<0.05$). The use of deferasirox among patients aged 13 to 17 years old was associated with higher SF-36v2 scores than in the other groups ($P<0.05$). However, for younger patients, the PedsQL scores were not significantly different for different iron chelators. The use of oral deferasirox significantly improved the quality of life of adolescents with b-thalassemia major. However, this effect was less prominent among patients aged 2 to 12 years old.

3. A study that carried out by (Musa, 2017) entitled Assessment The Quality of Life of Thalassemia Major Adolescents in Duhok City . A descriptive cross sectional study was conducted at Jeen pediatric hematology and oncology center in Duhok city for the period from Jan. 15th to May 25th. 2017 to assess quality of life of Adolescents suffering from Thalassemia Major, and finding a You mention to finding the relationship between domain but in result to applied differences between domains of quality of life and socio-demographic variables. Eighty-eight Adolescents randomly selected between ages (5–18 years) were enrolling in the study who is reviewing Jeen pediatric hematology and oncology center in Duhok city. Information were collected through the use Peds QL 4.0 generic core scale questionnaire, which included physical domain (8) items, emotional domain (5) items, social domain (5) items, and school domain (5) items. Results: The results of the study showed

that there were statistically significant correlation between the all domains of quality of life and some variables like education, family history, income level, frequency of blood transfusion, history of spleenectomy, and non-statistically significant correlation of some variables like age and gender , Conclusion: The quality of life scores were best for boys as compared to girls. Recommendations: The authorities should develop plans to improve quality of life of these patients. Whereas early diagnosis, regular visiting, financial and education support are the major factors of intervention to improve the overall quality of life of these Adolescents, the researcher also recommends conducting further studies on large samples.

4- A study (Evaluating Health-Related Quality of Life (HRQoL) in Iraqi Adult and Pediatric Patients with Beta-Thalassemia Major Using Two Different Iron Chelation Therapies) was conducted by (Awad, et al., 2019). A cross-sectional study was performed to evaluate health-related quality of life in both adult and pediatric patients using Deferoxamine (DFO) or Deferasirox (DFX) iron chelation therapies (ICTs) and to assess different factors affecting their elated quality of life. Two instruments were used to evaluate quality of life; the WHOQOL-BREF questionnaire was used to evaluate quality of life in adults (DFO group and DFX group), and PedsQL was used to evaluate quality of life in pediatrics (DFO group and DFX group). Several variables were assessed for correlation with quality of life domains. In adults patients; Health-Related Quality of Life (HRQoL) components were significantly lower in patients receiving Deferoxamine (DFO). In pediatric patients, only physical functioning score was significantly lower in Deferoxamine(DFO) receiving patients, the rest of the variables show no statistical differences. Pediatrics have slightly better quality of life (QoL) and lower complications in comparison to adults. Thalassemia impairs quality of life (QoL) in all patients significantly. Improving quality of life (QoL) is achieved by better control of iron overload, iron chelation therapies (ICTs) adverse effects, and complications..

5- It's done by (Odah et al., 2019) , entitled Hemoglobinopathies According To Blood Groups In Thi-Qar Governorate. In this cross- sectional study 601 patients with hemoglobinopathies (317 males and 284 females), Whose age ranged from months to ≥40 years. They were attending genetic blood disease center at Al- Habobi hospital in Thi-Qar governorate during the data collection period (1st March to 31 May 2016). Thalassemia has high prevalence than other hemoblobinopathies (76.53) and majority of cases was blood group O. The highly percentage of cases was at age ≤ 10 years (54.08) followed by 21-30 years was (34.44), highly percentage of cases was in urban (60.90). Adequate and suitable therapeutic and preventive measures should be taken to diminish birth of effected Adolescents such as health education of individuals about importance of premarital screening program for all couple to identify gen carrier individual of hemoglobinopathies.

Chapter Three

Methodology

This chapter provides description of steps that taken to conduct the present study. It contains research design, the administrative arrangement, settings of study, samples and data collection method, validity and study "reliability and also planning for analysis of data ".

3.1. study design:

A descriptive study was conducted on adolescent who have hemoglobinopathies at Hereditary Blood Disease Center in Karbala teaching Hospital for Children to assess the impact hemoglobinopathies on quality of life and adolescent behavior during the period 1st November /2020 to 3rd April/ 2022.

3.2. Administrative arrangement

Formal administrative approvals to perform the study were obtained prior to data collection, as shown in appendix (A) .

1.The first of all the researcher obtains an approval to the protocol of the study from the higher studies committee in College of Nursing at University of Babylon as seminar presentation, then viewing the protocol of the study to scientific research and ethical committee at the college and after that the protocol take the improvement.

2.The second steps after the protocol take the improvement from scientific research and ethical committee from co-dean of scientific affair and postgraduate and after that an official letter to facilitate the research mission directed to Directorate of Health in Karbala governorate.

3.Official approval was received from the Karbala health directorate and directed to the center for training and development in the Karbala health directorate, the primary approval took to facilitate the research mission and collect the data from hereditary blood disease center in Karbala teaching hospital for Adolescents, and after the meeting of research commitment of training and development center the final decision of Approval to conduct research had obtained.

3.3 .Ethical Considerations

Ethical obligations are one of the most important process that the investigator must follow when doing the study. Before the starting of collect the data from the community that has been identified for the study, the researcher should clarify the main purpose and desired goal of conducting this study for the sample to be including in the study, as well as adhere to the strict confidentiality of the data taken from the study sample and pledge to use it for scientific purposes related to the study only.

Before the starting of gathering the data from the sample who are participating in the study, the researcher given a brief explanation about the scientific background of the research and the purpose of conducting it and what is the role of the nurses who participate in this study, to give them a complete and clear idea about the study to be carried out. On the other hand, the researcher emphasized that all nurses who are participating in the study had the right to stop being part of and they can withdraw from this study in the event that they feel uncomfortable or annoyed with some of the items in the questionnaire that was prepared as a research tool, the researcher’s method of collecting data or anything else.

3.4. Setting of the study

The current study is carried out in the hereditary blood disease center in Karbala teaching hospital for Adolescents. This Center is chosen for the study because its the unique center in Karbala governorate .

3.5. The study sample:

A none–probability(convenient) sample of 200 hemoglobinopathies adolescent that selected from 256 adolescent who had admitted for management and registered in the heredity blood disease in Karbala city.

3.6. The instruments of the study

The instrument has been modify and adapted according to the purpose of the study .The questionnaire format is designed by:

1- Using specific scales to measure the variable of the present study quality of life questionnaire.

2- Using specific scales for measure Behaviors for Adolescent.

3- Review of the literatures and studies related to the concepts of the adolescent behavior and quality of life for adolescent.

3.7. Questionnaire format

It consists of the following parts(appendix(C):

A-Adolescent demographic data:

This section includes data about the adolescent age, gender, class, residency ,the birth order in the family…

B- Parent's demographic data

This part concerned with the parent’s demographic data; parent’s educational level, occupation, family income, family number, and residence …)

C-Adolescent clinical characteristics

This part consisted from the clinical data of the adolescent, age of the onset of illness, number of ill siblings ,number of blood transfusions ,clinical manifestations that displayed on the affected adolescent (such as chest pain ,joint pain ,loss of appetite ,spleen enlargement and heart disorders…..)

D- quality of life for adolescent

Quality of life assessment can be improved by using this tool because it meets various requirements. High validity and reliability have been proven in several studies on Adolescents with cancer, diabetes and heart disease, and is available in generic and for cancer, as well as for cerebral palsy, rheumatic heart disease, and thalassemia. It can be completed by Adolescents (Interview and self-report) or by parents/guardians (proxy report), has been translated into several languages for ease of use, and is available for a variety of age groups: 2-4 years (self and proxy report). James W. Varni, M.D., Ph.D., is the man credited with creating PedsQL. Since 2001, he's made a number of changes. There are multiple versions of this tool (from version 1.0 to 4.0), each with a different set of 23 questions spread throughout four different assessment areas: physical, emotional, social, and school. The 4.0 version is an all-purpose measurement tool. The PedsQL measures a Adolescents's overall well-being and can be used to compare chronically unwell Adolescents to healthy ones. Four domains of quality of life, comprising physical, emotional, social, and school well-being, were incorporated in this section's (23) questions:

1-physical domain:

A total of eight items are included (approximately). In order to walk, run, or engage in any other form of physical exercise, I have to be within 100 meters. It's tough for me to lift anything heavier than a certain amount. Thalassemia school-age Adolescents's physical health is manifested in a variety of ways, including difficulty bathing and showering alone, difficulty undertaking household chores, and low energy levels.

2- Emotional domain

There are a total of five components to this (I am worried or scared, I am sad or blue, I am angry, I am having difficulty sleeping, etc.). Because I don't know what's going to happen to me The disease's impact on the transmission of hemoglobinopathies in teenagers is depicted in all of these ways.

3-Social domain

Five things are included. (I have a hard time getting along with other youngsters,ect.). Hemoglobinopathies adolescent Adolescents have a difficult time relating to other teenagers because of their condition.

4-School functioning:

There are five parts to it: Adolescents with hemoglobinopathies may find it difficult to pay attention in class, forget things, and struggle to keep up with their schoolwork due to the disease's affects on their education. Similarly, they often skip school due to illness or to visit a doctor or hospital.

Part Four: Adolescent behavior scale:

Adolescent Behavior Checklist (ABC) is designed to assess characteristic of behavior in adolescents between the ages of 11 and 17 years. The ABC consists of 48 items, 42 of which yield a total ABC score and six factor scores (Conduct Problems, Impulsivity/Hyperactivity, Poor Work Habits, Inattention, Emotional Lability, Social Problems). Adolescent respondents are instructed to rate each item on a 4- point Likert scale "not at all" (0), "just a little" (1), "pretty much" (2), and "very much" (3).

3.8. Rating and Scoring of the Study Instrument:

The quality of life questionnaire form has been scored and rated on five levels Likert scale (0) points for never, (1) points for Almost never ,(2) point sometimes , (3) point for often , and (4) point for Almost always which were evaluated from a cutoff point (٢) based on scores (0,1, 2, 3and 4), respectively.

The adolescent behavior questionnaire form has been scored and rated at four levels Likert scale, (0) point for not at all, (1) point for just a little, (2) point for pretty much, and (3) point for very much respectively.

3.9. Validity of the instrument

The validity: The extent to which the research tool measures what it is supposed to measure has been characterized as the validity of the instrument. The tool should cover all aspects of the subject under investigation (Polit and Hungler , 2013).

Face validity, this is a simple form of validity that essentially confirms that the instrument appears to be measuring the idea. In an intuitive type of validity, participants or coworkers are asked to read the instrument and assess if the material seems to match the concept that the researcher is trying to measure (Haber and LoBiondo-Wood, 2014).

These experts were requested to examine the questionnaire for content clarity, relevance, and appropriateness; their responses revealed that a few items needed minor revisions. These changes were made in response to their recommendations and insightful comments (Appendix B).

3.10.Pilot study

A small-scale investigation known as a pilot study was carried out between the period (1st) of March 2021 to (15) of March 2021, The pilot study was carried out hereditary blood disease in Karbala teaching hospital for Adolescents., the researcher was selected (10%) of total sample that about (20) adolescent;, the instrument filled out by them and after that reliability was achieved. The researcher conducted the pilot study to:

1. Assess the items accuracy, relevancy and effectiveness of assessing tool.
2. Determine the period of time that needs for collecting data.
3. Testing the study tool' reliability.
4. Identify the obstacles that may be facing the researcher throughout the data gathering procedure.

3.11. Reliability

The reliability of the items of the questionnaire format determined by using of the internal consistency .

The scope by which a consistently measures of concepts throughout an instrument was termed as reliability (Burns& Grove 2010). The purpose of the pilot study was to determine the research tool's reliability. In order to determine the study instrument's reliability, Cronbach's Alpha was determined using the IBM

SPSS version 25.0 statistical package. The instrument's reliability was (r = 0.843, 0.711, 0.762)

The result of the Cronbach's Alpha reliability of the study tool was statistically high, indicate that the study tool was reliable and has equal measurability.

Table (3-1) indicate this instrument has good reliability .

N. items	Reliability technique	Actual values
Quality of life 23 items	**Cronbach's Alpha**	**0.835**
Behavior scale	**Cronbach's Alpha**	**0.862**

3.12 Data collection method

The data collected from March 2021 and July 2021. By conducting a focused interview with each subject, the researcher was able to gather information about the study's factors. Data acquired from patients at Heredity Blood Diseases in Karbala Teaching Hospital for Adolescents. A questionnaire-based interview takes about 20 to 25 minutes per subject. The verbal agreement to participate in the study is secured. Participants were informed that the study was ethically sound and that any information they submitted would be treated with the strictest of confidence.

3.13.Statistical Analysis

The package of SPSS version 20 is used to analyze the data through the application of statistical procedures as following:

3.13.1. Descriptive statistical methods

The Frequency (f) Percentage (percent) =,f/n100 is used to determine this method.

3.13.2.Inferential statistical procedures

3.13.2.a Analysis of Variance" (ANOVA): ANOVAs are used for comparing (testing) three or more means for statistical significance. They are statistical models used to investigate variations among group means and their associated procedures (such as "variance between and among groups"). While this test is similar in idea to multiple two-sample t-tests, it is less conservative (resulting in less type I error).), making it useful for a wider range of applications (Fisher & Yates, 2016). We looked at the connections between nurses' demographic data such as their age, educational level, marital status, place of employment and number of years on the job as well as their training course.

3.13.2.b. Independent t-test: An inferential statistical test known as the two sample t-test, independent-sample t-test, or student's t test is used to evaluate whether there is a statistically significant difference in mean values between unrelated groups. (According to Leard Statistics, 2019). It was utilized to find a link between nurses' levels of knowledge, attitudes, and practices and their demographic factors, such as gender and residency. It has the following formula:

3.13.2.c. Correlation coefficient:

An interval or ratio scale can be used to determine the link between two variables , and the Pearson correlation coefficient is a parametric test that can do just that. Pain management knowledge, attitudes, and practices among nurses are highly correlated, and this information can be used to identify any positive or negative relationships. At a significance level of, the P-value is found to be statistically significant (0.05)

Chapter Four
Results of the Study

This chapter extensively introduces the outcomes of the research in tables and these refer to the objectives of this report, which are as follows:

4.1.Sample Characteristics

Table4-1-1.Demographic information of Adolescents

Factors	Classification	Freq.=200	%
Age/years	11-13years old	60	30.0
	14-16years old	76	38.0
	17 and more	64	32.0
	Total	200	100.0
Gender	Male	91	45.5
	Female	109	54.5
	Total	200	100.0
Education level	Not read and write	3	1.5
	Read and write	46	23.0
	Primary school	116	58.0
	High school	35	17.5
	Total	200	100.0
Number of sibling	No	17	8.5
	1	145	72.5
	>1	38	19.0
	Total	200	100.0
Order	1st	46	23.0
	2nd	144	72.0
	3rd and above	10	5.0
	Total	200	100.0

Findings show participants age, the age 14-16 years old records the highest percentage (38%). In regards with gender, female adolescents were predominated (54.5%). Also the level of education the findings of studied sample are (58%) primary school adolescents. In terms of number of sibling, most of students

(72.5%) were had 1 sibling. It is obvious form the findings (72%) that the second order adolescents within their family.

Table4-1-2.Clinical information of Adolescents

Factors	Classification	Freq.=200	%
Disease related relatives	No	48	24.0
	1st degree	51	25.5
	2nd degree	45	22.5
	3rd degree	56	28.0
	Total	200	100.0
Transfusion therapy	Regular	102	51.0
	Irregular	98	49.0
	Total	200	100.0
Iron chelation therapy	Regular	137	68.5
	Irregular	63	31.5
	Total	200	100.0
Number of transfusion therapy	Once time per month	64	32.0
	Twice time per month	136	68.0
	Total	200	100.0

In terms of frequencies and percentage, adolescents (28.5%) exhibit a first degree of disease related relative. Transfusion therapy related findings the studied sample (51%) were regular therapy. In terms of iron chelation therapy, most of students (68.5%) were regular therapy. It is obvious form the findings (68%) that the twice transfusion therapy.

Table4-1-3.Socio-demographic Characteristic of Adolescents Family ?

Factors	Classification	Freq.=200	%
Fathers Occupation	Employ	89	44.5
	Free work	47	23.5
	Retired	3	1.5
	Unemployment	61	30.5
	Total	200	100.0
Mothers Occupation	Employ	67	33.5
	Free work	4	2.0
	Retired	120	60.0
	Housewife	9	4.5
	Total	200	100.0
Fathers Education	Not read and write	13	6.5
	Read and Write	44	22.0
	Primary School	33	16.5
	High School Graduate	54	27.0
	Institute or College	56	28.0
	Total	200	100.0
Mothers Education	Not read and write	10	5.0
	Read and Write	47	23.5
	Primary School	46	23.0
	High School Graduate	54	27.0
	Institute or College	43	21.5
	Total	200	100.0
Family type	Nuclear	157	78.5
	Extended	43	21.5
	Total	200	100.0
Residents	Rural	180	90.0
	Urban	20	10.0
	Total	200	100.0
Economic status	Insufficient	19	9.5
	Slightly sufficient	104	52.0
	Sufficient	77	38.5
	Total	200	100.0

From the findings that the (44.5% and 33%) of parents were employed. In regards with education, most of parents were institute and college graduated. The families, (78.5%) with nuclear families. The study sample concerned with residents (rural) 90% and the economic status were (52%)slightly sufficient..

4.2.Quality of Life for Hemoglobinopathies Adolescent

Table4-2-1.Quality of Life related to Physical Functioning

List	Physical Function Items	Responses	Freq.	%	*M ± SD*	*Ass.*
1	It is hard for me to walk more than 100M	Almost Always	53	26.5	*2.14±1.419*	*Moderate*
		Often	2	1.0		
		Sometime	35	17.5		
		Almost Never	83	41.5		
		Never	27	13.5		
2	It is hard for me to run	Almost Always	64	32.0	*2.01±1.463*	*Moderate*
		Often	3	1.5		
		Sometime	17	8.5		
		Almost Never	99	49.5		
		Never	17	8.5		
3	It is hard for me to do sports activity or exercise	Almost Always	55	27.5	*2.15±1.425*	*Moderate*
		Often	1	.5		
		Sometime	25	12.5		
		Almost Never	96	48.0		
		Never	23	11.5		
4	I find it difficult to lift anything substantial.	Almost Always	77	38.5	*1.75±1.462*	*Moderate*
		Often	32	16.0		
		Sometime	78	39.0		
		Almost Never	13	6.5		
		Never	0	0.0		
5	It is hard for me to take a bath or shower by myself	Almost Always	36	18.0	*2.65±1.462*	*Moderate*
		Often	1	.5		
		Sometime	41	20.5		
		Almost Never	41	20.5		
		Never	81	40.5		
6	Doing the duties around the house is difficult for me.	Almost Always	41	20.5	*2.33±1.353*	*Moderate*
		Often	1	.5		
		Sometime	44	22.0		
		Almost Never	78	39.0		
		Never	36	18.0		
7	I hurt or ache	Almost Always	63	31.5	*1.83±1.329*	*Moderate*
		Often	2	1.0		
		Sometime	44	22.0		
		Almost Never	87	43.5		
		Never	4	2.0		
8	I have low energy	Almost Always	83	41.5	*1.69±1.467*	*Moderate*
		Often	19	9.5		
		Sometime	92	46.0		
		Almost Never	6	3.0		
		Never	0	0.0		

(M.s) Mean of scores, (SD) Standard deviation
(Poor= 0-1.33; Moderate=1.34-2.66; Good= 2.67-4)

This table show it is the mean score and standard deviations of the items reveal that adolescent experience moderate physical function as indicated by moderate mean scores all items .

Table4-2-2:Overall Quality of Life related to Physical Functioning among Hemoglobinopathies Adolescent

Physical Function	Freq.	%	*M ± SD*
Poor (*M=0-10*)	45	22.5	*16.57±8.62*
Moderate (*M=11-21*)	85	42.5	
Good (*M=22-32*)	70	35.0	
Total	200	100.00	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of physical function related to quality of life was demonstrate that (42.5%) of adolescent are experienced moderate with average of *M ± SD=16.57±8.62*.

Table4-2-3.Quality of Life related to Emotional Functioning

List	Emotional Functioning Items	Responses	Freq.	%	*M ± SD*	*Ass.*
1	I feel afraid or scared	Almost Always	67	33.5	*1.97±1.546*	*Moderate*
		Often	7	3.5		
		Sometime	24	12.0		
		Almost Never	69	34.5		
		Never	33	16.5		
2	I feel sad or blue	Almost Always	75	37.5	*1.87±1.566*	*Moderate*
		Often	8	4.0		
		Sometime	11	5.5		
		Almost Never	80	40.0		
		Never	26	13.0		
3	I feel angry	Almost Always	70	35.0	*1.79±1.474*	*Moderate*
		Often	12	6.0		
		Sometime	26	13.0		
		Almost Never	73	36.5		
		Never	19	9.5		
4	I have trouble sleeping	Almost Always	89	44.5	*1.56±1.532*	*Moderate*
		Often	9	4.5		
		Sometime	22	11.0		
		Almost Never	60	30.0		
		Never	20	10.0		
5	When I think of the future, I am afraid of what might happen to me.	Almost Always	52	26.0	*2.29±1.606*	*Moderate*
		Often	9	4.5		
		Sometime	38	19.0		
		Almost Never	30	15.0		
		Never	71	35.5		

(M.s) Mean of scores, (SD) Standard deviation
(Poor= 0-1.33; Moderate=1.34-2.66; Good= 2.67-4)

This table show it is the mean score and standard deviations of the items reveal that adolescent experience moderate emotional function as indicated by moderate mean scores among all items .

Table4-2-4:Overall Quality of Life related to Emotional Functioning among Hemoglobinopathies Adolescent

Emotional Functioning	Freq.	%	*M* ± *SD*
Poor (*M=0-6*)	63	31.5	*9.49±6.55*
Moderate (*M=7-13*)	73	36.5	
Good (*M=14-20*)	64	32.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of emotional function related to quality of life was demonstrate that (36.5%) of adolescent are experienced moderate with average of *M* ± *SD=9.49±6.55*.

Table4-2-5.Quality of Life related to Social Functioning

List	Social Functioning Items	Responses	Freq.	%	*M ± SD*	*Ass.*
1	I have trouble getting along with other Adolescentss	Almost Always	72	36.0	*1.85±1.521*	*Moderate*
		Often	3	1.5		
		Sometime	35	17.5		
		Almost Never	62	31.0		
		Never	28	14.0		
2	Other teenagers do not want to be my friend	Almost Always	33	16.5	*2.55±1.423*	*Moderate*
		Often	4	2.0		
		Sometime	57	28.5		
		Almost Never	32	16.0		
		Never	74	37.0		
3	Other teenagers tease me	Almost Always	36	18.0	*2.33±1.319*	*Moderate*
		Often	4	2.0		
		Sometime	56	28.0		
		Almost Never	65	32.5		
		Never	39	19.5		
4	Things that most teenagers my age can accomplish come naturally to me don't.	Almost Always	55	27.5	*2.01±1.398*	*Moderate*
		Often	5	2.5		
		Sometime	45	22.5		
		Almost Never	72	36.0		
		Never	23	11.5		
5	With other teenagers, keeping up is a real challenge.	Almost Always	67	33.5	*1.90±1.475*	*Moderate*
		Often	3	1.5		
		Sometime	35	17.5		
		Almost Never	72	36.0		
		Never	23	11.5		

(M.s) Mean of scores, (SD) Standard deviation
(Poor= 0-1.33; Moderate=1.34-2.66; Good= 2.67-4)

This table show it is the mean score and standard deviations of the items reveals that adolescent are experienced moderate social function as indicated by moderate mean scores among all items .

Table4-2-6:Overall Quality of Life related to Social Functioning among Hemoglobinopathies Adolescent

Social Functioning	Freq.	%	*M* ± *SD*
Poor (*M=0-6*)	42	21.0	*10.66±5.98*
Moderate (*M=7-13*)	84	42.0	
Good (*M=14-20*)	74	37.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of social functioning related to quality of life was demonstrate that (42%) of adolescent are experienced moderate with average of *M* ± *SD=10.66±5.98*.

Table4-2-7.Quality of Life related to School Functioning

List	School Functioning Items	Responses	Freq.	%	*M ± SD*	*Ass.*
1	. In school, it's difficult to pay attention.	Almost Always	62	31.0	*2.11±1.576*	*Moderate*
		Often	7	3.5		
		Sometime	21	10.5		
		Almost Never	66	33.0		
		Never	44	22.0		
2	I forget things	Almost Always	51	25.5	*2.12±1.492*	*Moderate*
		Often	17	8.5		
		Sometime	30	15.0		
		Almost Never	61	30.5		
		Never	41	20.5		
3	To keep up with my academics, I'm having a hard time	Almost Always	59	29.5	*2.09±1.547*	*Moderate*
		Often	6	3.0		
		Sometime	40	20.0		
		Almost Never	48	24.0		
		Never	47	23.5		
4	Because I'm sick, I'm unable to go to school.	Almost Always	42	21.0	*2.33±1.501*	*Moderate*
		Often	9	4.5		
		Sometime	56	28.0		
		Almost Never	26	13.0		
		Never	67	33.5		
5	To go to the doctor or hospital, I have to miss school.	Almost Always	47	23.5	*2.25± 1.475*	*Moderate*
		Often	8	4.0		
		Sometime	42	21.0		
		Almost Never	54	27.0		
		Never	49	24.5		

(M.s) Mean of scores, (SD) Standard deviation
(Poor= 0-1.33; Moderate=1.34-2.66; Good= 2.67-4)

This table show it is the mean score and standard deviations of the items reveals that adolescent are experienced moderate school function as indicated by moderate mean scores among all items.

Table4-2-8:Overall Quality of Life related to School Functioning among Hemoglobinopathies Adolescent

School Functioning	Freq.	%	*M ± SD*
Poor (*M=0-6*)	44	22.0	*10.91±5.81*
Moderate (*M=7-13*)	86	43.0	
Good (*M=14-20*)	70	35.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of school functioning related to quality of life was demonstrate that (43%) of adolescent are experienced moderate with average of *M ± SD=10.91±5.81*.

Table4-2-9:Overall Quality of Life related among Hemoglobinopathies Adolescent

Quality of Life	Freq.	%	*M ± SD*
Poor (*M=0-42*)	71	35.5	*47.63±24.38*
Moderate (*M=43-85*)	121	60.5	
Good (*M=86-128*)	8	4.0	
Total	200	100.0	

M: Mean for total score, SD=Standard Deviation for total score

The analysis of quality of life among Hemoglobinopathies adolescents was demonstrate that (60.5%) are experienced moderate quality of life with average of *M ± SD=47.63±24.38*.

Figure4-5:Quality of Life among Hemoglobinopathies Adolescent

Table4.3.Health Behaviour among Hemoglobinopathies Adolescent

List	Health Behaviour Items	*M ± SD*	*Ass.*
1	I have difficulty sitting still	*1.82±0.887*	*Poor*
2	I get excited easily	*2.11±0.933*	*Moderate*
3	I say or do things without thinking.	*2.13±0.999*	*Moderate*
4	I have trouble concentrating.	*2.12±1.051*	*Moderate*
5	I get frustrated easily.	*2.28±1.076*	*Moderate*
6	I get angry easily.	*2.1±1.060*	*Moderate*
7	I get into trouble with adults, such as principals, teachers, employers. dangerous things without thinking what might happen.	*2.14±0.989*	*Moderate*
8	I argue with my family members.	*1.98±1.056*	*Poor*
9	I argue with people outside my family.	*2.60±1.177*	*Moderate*
10	My moods change quickly.	*2.10±1.219*	*Moderate*
11	I purposely break rules or disobey instructions.	*1.83±1.069*	*Moderate*
12	I do dangerous things without thinking what might happen.	*2.32±0.981*	*Moderate*
13	I’m afraid of losing control or becoming violent.	*2.39±1.120*	*Moderate*
14	I am stubborn or strong-willed.	*1.35±0.861*	*Poor*
15	I get into trouble with the police; I break laws.	*1.57±0.948*	*Poor*
16	I steal things.	*1.95±1.021*	*Poor*
17	I lie to people.	*1.89±1.016*	*Poor*
18	I skip school; I am truant.	*2.10±1.014*	*Moderate*
19	I am disorganized.	*2.28±1.003*	*Moderate*
20	I have trouble making or keeping friends.	*1.81±0.804*	*Moderate*
21	My mind wanders easily; I daydream.	*2.04±1.048*	*Moderate*
22	I am easily distracted by noises.	*2.19±0.965*	*Moderate*
23	I have trouble staying seated.	*2.10±0.958*	*Moderate*
24	I have problems waiting my turn in games or group situations.	*2.03±1.038*	*Moderate*
25	I forget to do my chores or other things I'm asked to do.	*2.70±1.056*	*Moderate*
26	I often blurt out answers to questions before they are completed.	*2.55±1.087*	*Moderate*
27	I talk too much.	*2.26±1.063*	*Moderate*
28	I feel restless and nervous.	*2.01±1.004*	*Moderate*
29	I cry easily.	*1.64±0.801*	*Poor*
30	I have difficulty learning in school.	*2.15±0.970*	*Moderate*
31	I have problems paying attention to instructions.	*1.99±0.879*	*Poor*
32	I interrupt others when they are speaking.	*1.66±0.957*	*Poor*
33	I lose things necessary to do my classwork (e.g., pencil, books).	*1.66±0.898*	*Poor*
34	I don’t complete my classwork.	*1.45±0.800*	*Poor*
35	I’m physically cruel to animals or people.	*1.57±0.668*	*Poor*
36	I swear or use bad language.	*1.94±0.839*	*Poor*
37	I disturb or annoy other people.	*2.13±1.082*	*Moderate*
38	I blame others for my own mistakes.	*2.11±0.946*	*Moderate*
39	I get into physical fights.	*2.01±1.022*	*Moderate*
40	I waste time when studying.	*1.73±1.011*	*Poor*
41	Most of my friends are younger than I am.	*1.71±0.942*	*Poor*
42	1 have problems remembering what I read.	*1.92±0.918*	*Poor*
43	My friends get annoyed with me.	*2.05±0.998*	*Moderate*

44	I don't get along well with peers of the opposite sex.	*1.80±0.885*	*Poor*
45	I don't complete my homework.	*1.77±1.004*	*Poor*
46	I make careless errors in my classwork or homework.	*2.07±1.017*	*Moderate*
47	My classwork or homework is sloppy.	*1.77±0.718*	*Poor*

"(M) Mean, (SD) Standard deviation, Level of Assessment (Poor= 1-2, Moderate= 2.1 – 3, Good= 3.1 – 4)"

This table show it is the mean score and standard deviations of the items reveals that adolescent are experienced poor level (*M> 2*) as indicated by low level of mean scores to moderate level (*M=2-3*) as described by moderate mean scores of the health behaviour at items of the scale.

Table4-3-2:Overall Health Behaviour among Hemoglobinopathies Adolescent

Health Behaviour	Freq.	%	*M ± SD*
Poor (*M=47-94*)	98	49.0	*94.0±28.74*
Moderate (*M=95-141*)	82	41.0	
Good (*M=142-188*)	20	10.0	
Total	200	100.00	

M: Mean for total score, SD=Standard Deviation for total score

Findings demonstrated that the (49%) of Hemoglobinopathies adolescents were poor health behaviour with average of *M ± SD=94.0±28.74*.

4.4. Differences in Quality of Life and Health Behaviour with regards Adolescents Information

Table 4-4-1: Differences in QoL and Health Behaviour with regard Adolescents Age (*n=200*)

Age	Source of variance	Sum of Squares	d.f	Mean Square	F	*p≤ 0.05*
QoL	Between Groups	9.293	2	4.647	4.270	.015
	Within Groups	214.348	197	1.088		
	Total	223.642	199			
Health Behavior	Between Groups	.540	2	.270	.734	.481
	Within Groups	72.494	197	.368		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

Quality of life differed significantly between age groups, although there were no significant variations in health behavior among teenagers with hemoglobinopathies (p>0.05).

Table 4-4-2:Significant Differences in QoL and Health Behaviour with regard Adolescents Gender (*n=200*)

Variables	Gender	Mean	SD	t-test	D.F	*p≤ 0.05*
QoL	female	2.4262	.86419	4.536	198	*.000*
	Male	1.7746	1.11958			
Health behaviour	Male	2.0692	.61879	1.469	198	*.143*
	Female	1.9432	.59149			

d.f: Degree of freedom, F: F-statistic

Health behavior among hemoglobinopathies teenagers did not show any significant differences in gender (p>0.05), however there were significant disparities in quality of life among men and women (p0.05).

Table 4-4-3: Differences in QoL and Health Behaviour with regard Adolescents Education Level (*n=200*)

Education level	Source of variance	The sum of the squares.	D.F	Mean of Square	F	*p-value*
QoL	Within Groups	3.662	3	1.221	1.088	.355
	Between Groups	219.980	196	1.122		
	Total	223.642	199			
Health Behavior	Between Groups	.607	3	.202	.547	.651
	Within Groups	72.427	196	.370		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

For hemoglobinopathies teenagers, there were no significant variations in quality of life and health behavior (p>0.05).

Table 4-4-4:Significant Differences in QoL and Health Behaviour with regard Adolescents Number of Sibling (*n=200*)

No. sibling	Source of variance	Sum of Squares	d.f	Mean Square	F	*p≤ 0.05*
QoL	Between Groups	3.969	2	1.985	1.780	.171
	Within Groups	219.672	197	1.115		
	Total	223.642	199			
Health Behavior	Between Groups	.156	2	.078	.211	.810
	Within Groups	72.877	197	.370		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

Results showed there were no significant variations in quality of life and health behavior among adolescents with hemoglobinopathies (p>0.05) based on their number of siblings.

Table 4-4-5: Differences in QoL and Health Behaviour with regard Adolescents Order of Adolescents (*n=200*)

Order	the source of variation	The sum of the squares.	D.F	Square Root of Mean	F	*P-value*
QoL	Within Groups	.716	2	.358	.316	.729
	Between Groups	222.926	197	1.132		
	Total	223.642	199			
Health Behavior	Between Groups	.453	2	.227	.615	.542
	Within Groups	72.580	197	.368		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

Findings showed that teenagers with hemoglobinopathies in their family had no significant variations in quality of life and health behavior (p>0.05).

Table 4-4-6:Significant Differences in QoL and Health Behaviour with regard Disease Related Relives (*n=200*)

Disease related relives	the source of variation	The total of the squares.	D.F	Squared Mean	F	*Sig.*
QoL	Within Groups	9.921	3	3.307	3.033	.030
	Between Groups	213.721	196	1.090		
	Total	223.642	199			
Health Behavior	Between Groups	3.781	3	1.260	3.567	.015
	Within Groups	69.252	196	.353		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

In the study of adolescents with hemoglobinopathies, there were significant variations in health-related quality of life and behavior (p0.05).

Table 4-4-7:Differences in QoL and Health Behaviour with regard Transfusion Therapy (*n=200*)

Transfusion therapy	Source of variance	Total of Squares	D.F	Squared Mean	F	*Sig.*
QoL	Within Groups	.220	1	.220	.195	.659
	Between Groups	223.422	198	1.128		
	Total	223.642	199			
Health Behavior	Between Groups	.101	1	.101	.274	.601
	Within Groups	72.933	198	.368		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

Findings showed that there were no significant variations in quality of life and health behavior among adolescents with hemoglobinopathies who received transfer therapy ($p>0.05$).

Table 4-4-8:Significant Differences in QoL and Health Behaviour with regard Iron Chelation Therapy (*n=200*)

Iron chelation therapy	Source of variance	Total of Squares	D.F	Square Root of Mean	F	*Sig.*
QoL	Within Groups	3.761	1	3.761	3.386	.067
	Between Groups	219.881	198	1.111		
	Total	223.642	199			
Health Behavior	Between Groups	.170	1	.170	.463	.497
	Within Groups	72.863	198	.368		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

The results showed that there were no significant differences in quality of life or health behavior among hemoglobinopathies teenagers in relation to iron chelation therapy (p0.05).

Table 4-4-9:Differences in QoL and Health Behaviour with regard Number of Transfusion Therapy (*n=200*)

No. transfusion therapy	Source of variance	Total of Squares	D.F	Square Root of Mean	F	*Sig.*
QoL	Within Groups	1.689	2	.845	.750	.474
	Between Groups	221.952	197	1.127		
	Total	223.642	199			
Health Behavior	Between Groups	.164	2	.082	.222	.801
	Within Groups	72.869	197	.370		
	Total	73.033	199			

d.f: Degree of freedom, F: F-statistic

There were no significant variations in quality of life or health behavior amongst hemoglobinopathies teenagers (p0.05) when it came to the amount of transfusions they received.

4.5.Correlation between Quality of Life and Health Behaviour with regard to Hemoglobinopathies Adolescents their Parents Socio-Demographic Information

Table4-5-1:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Fathers Occupation

	Fathers Occupation		Sig.
QoL	*Sig. (2-tailed)*	*.155**	Positive Sig.
	Pearson's r	*.028*	
Health Behaviour	*Sig. (2-tailed)*	*.112*	Non-sig.
	Pearson's r	*.116*	

Findings exhibit there were significant correlation (positive) between quality of life of hemoglobinopathies adolescents and fathers occupation (*r=0.155; p=0.028*).

Table4-5-2:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Mothers Occupation

	Mothers Occupation		Sig.
QoL	*Pearson's r*	*.126*	No-sig.
	Sig. (2-tailed)	*.076*	
Health Behaviour	*Pearson's r*	*- .243***	Negative-Sig.
	Sig. (2-tailed)	*.001*	

Findings exhibit there were significant correlation (negative) between health behaviour of hemoglobinopathies adolescents and mothers occupation (*r= -0.243; p=0.001*).

Table4-5-3:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Fathers Education

	Fathers Education		Sig.
QoL	*Sig. (2-tailed)*	*.124*	Non-sig.
	Pearson's r	*.081*	
Health Behaviour	*Sig. (2-tailed)*	*.097*	Non-sig.
	Pearson's r	*.172*	

Findings exhibit there were no-significant correlation between quality of life and health behaviour of hemoglobinopathies adolescents with regards fathers education ($p>0.05$).

Table4-5-4:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Mothers Education

	Mothers Education		Sig.
QoL	*Sig. (2-tailed)*	*.161**	Positive-Sig.
	Pearson's r	*.022*	
Health Behaviour	*Sig. (2-tailed)*	*.091*	Non-sig.
	Pearson's r	*.198*	

Findings exhibit there were significant correlation (positive) between quality of life of hemoglobinopathies adolescents and mothers education ($r= -0.161; p=0.022$).

Table4-5-5:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Family Type

	Family Type		Sig.
QoL	*Sig. (2-tailed)*	*.012*	Non-sig.
	Pearson's r	*.864*	
Health Behaviour	*Sig. (2-tailed)*	*.143**	Positive-Sig.
	Pearson's r	*.044*	

Findings exhibit there were significant correlation (positive) between health behaviour of hemoglobinopathies adolescents with regards family type (*r= -0.143; p=0.044*).

Table4-5-6:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Residences

	Residents		Sig.
QoL	*Sig. (2-tailed)*	*- .029*	No-sig.
	Pearson's r	*.685*	
Health Behaviour	*Sig. (2-tailed)*	*- .006*	Non-sig.
	Sig. (2-tailed)	*.928*	

Findings exhibit there were no-significant correlation between quality of life and health behaviour of hemoglobinopathies adolescents with regards residents (*p>0.05*).

Table4-5-7:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Family Economic

	Economic Status		Sig.
QoL	*Sig. (2-tailed)*	*0.252***	Positive-Sig.
	Pearson's r	*0.000*	
Health Behaviour	*Sig. (2-tailed)*	*.188**	Positive-Sig.
	Pearson's r	*.008*	

The quality of life and health behavior of teens with hemoglobinopathies were strongly linked to their parents' economic status (r=0.252; p=0.000; and r=0.188; p=0.008 respectively), and this correlation was very significant (positive).

Table 4-5.8 Correlation Between Quality of Life and Health Behaviour among Hemoglobinopathies Adolescents (*n=200*)

Health Behaviour			
QoL	*Spearman's rho*	*.260**	Positive Sig.
	Sig. (2-tailed)	*.000*	
	N	*400*	

Findings exhibit there were significant correlation (positive) between health behaviour and quality of life of hemoglobinopathies adolescents (*r=0.260; p=0.000*).

Chapter Five
Discussion of the Results

Table4-1-1.Demographic information of Adolescents

Findings show participants age, the age 14-16 years old records the highest percentage . In regards with gender, female adolescents were predominated more than half. Education level related findings, more than half of studied sample more than half were primary school adolescents. In terms of number of sibling, most of students more than two third were had 1 sibling. It is obvious form the findings more than two third that the second order adolescents within their family.

Concerning age , and level of education the finding of present study consistent with (Shaker and Nasir, 2013) who carried out study to assess thalassemia adolescents′ quality of life through it domain of school functioning, and found that the majority of the study sample fall within the second category of age (12- 13yrs), and the majority of them are primary school graduated.

Regarding gender, this finding incongruent with (Musa, 2017) who conducted a study about Assessment the Quality of Life of Thalassemia Major Adolescents in Duhok City, and found that the most of sample were male. This differences may be due to sampling selection, and culture disparities.

In respect to number of sibling, this result in congruent with (Mansoor et al., 2018) who carried out a study , and illustrated that more than half of sample have 1-2 sibling.

Regarding Adolescents order, this finding inconsistent with (El-Said Zaghamir et al., 2019) who who reported that most of sample were first birth order, the researcher believed that differences may be due to natural of Iraq society , and may be study sample differences.

Table4-1-2.Clinical information of Adolescents

In terms of frequencies and percentage, adolescents one quarter exhibit a first degree of disease related relative. Transfusion therapy related findings, half of studied sample more than half were regular therapy. In terms of iron chelation therapy, most of students more than two third were regular therapy. It is obvious form the findings more than two third that the twice transfusion therapy.

Regarding chelation therapy, this finding in the same line with (Ayoub et al., 2013) who found that most of sample receive chelation therapy. This may be due to the drug choice for removal of iron excess due to the increased production of immature RBCs and their rapid destruction .

According to (Mansoor et al., 2018), more than half of the sample having blood transfusion therapy had their results confirmed. Every two weeks or so, on average

This finding is consistent with (Mahmoud, 2019) who conducted a study on the quality of life of Adolescents with thalassemia in Alexandria and discovered that consanguinity was detected among more than half of the parents. This may be due to the consanguinity major cause of thalassemia.

Table4-1-3.Socio-demographic Characteristic of Adolescents Family

From the findings that the highest percentage of parents were employed. In regards with education, most of parents were institute and college graduated. Most of families, more than two third nuclear families. The majority of study sample were rural residents. More than half were slightly sufficient economic status.

Regarding occupation, this finding consistent with study entitled (Psycho-social Development of Thalassemia and Healthy Adolescents: Comparative Study) that carried by (Saeed and aldoori, 2020) , who reported that the most of sample were employed.

Concerning level of education, this finding congruent with study that carried out by (Ali, 2018) who stated that highest percentage of mother studies university.

Regarding type of family, this finding go along with study that carried out by (Mathew, 2018) who reported that most of family belong to nuclear family. This may be due to nature of Iraqi society culture, and traditional.

Concerning resident, the result of current study in similarity with study that carried out by (Essawy et al., 2018) who illustrated that most of sample were living in rural area. This may be due to the major cause of thalassemia the marriage from relatives, and this is according to the habits and culture of Iraqi society.

Regarding economic status, this finding corresponding with study that conducted by (Tuama et al., 2015) who found that half of sample the income Somewhat Sufficient. This outcome is the product of numerous adjustments made by private clinics, doctors, and pharmaceutical companies, as well as the unique requirements of individual patients. This variance may belong to the contentment that widely spreading in Iraqi community and that did not indicated in necessary they are with good socio-economic status but it give them sense of satisfying.

4.2.Quality of Life for Hemoglobinopathies Adolescent

Table4-2-1.Quality of Life related to Physical Functioning

The mean score and standard deviations of the items reveals that adolescent are experienced moderate physical function as indicated by moderate mean scores among all items except, the items number (4, 7 and 8) the responses were poor physical function as indicated by low level of mean.

According to a cross-sectional study (Nashwan et al., 2018) that assessed the quality of life among patients aged 14 to 18 years with thalassemia major (TM), the results indicated that Adolescents with the disease had a significantly lower and more variable overall quality of life score (69.1–16.8) than healthy controls.

According to the researcher's opinion, this may be due to, the thalassemia effect on the Adolescents, in all aspects of life including social, emotional, and physical.

In the same direction, Hassan et al., 2019 who conducted a study about Study of the Health Instructions Effect on Quality of Life and Psychological Problems among Adolescents with Thalassemia, and reported that, more than two third of patients more than half scored medium grade regarding their physical functioning. This may be due to, the thalassemia disease effect on the circulation of the Adolescents, and cause decreased hemoglobin that considers necessary for carrying oxygen.

The decreased physical activity may be imputed to the effects of thalassemia on the body skeleton which results in low bone sizes and delayed puberty .The rapid fatigue and low physical may be also caused by cardiovascular disorders that are known to be present in many adolescents with thalassemia .

Table4-2-2:Overall Quality of Life related to Physical Functioning among Hemoglobinopathies Adolescent

The analysis of physical function related to quality of life was demonstrate that highest percentage of adolescent are experienced moderate with average of *M ± SD=16.57±8.62*.

This finding in the same line with study that carried out by (Mahmoud, 2019) who reported that Nearly three quarters of the Adolescents more than two third had ”low” QOL regarding physical functioning.

In the same context, (Ankush et al., 2019) who conducted a study to Quality of life in Adolescents with thalassemia major following up at a tertiary care center in India (GOTQoL) , and found that the Adolescents with thalassemia had lower mean scores on physical due to the frequency of blood transfusion and iron chelation, the physical decline .

Anemia and other problems of iron overload may be to blame for the discovery, which is confirmed by the evidence (Cheuk et al, 2008). the normal time of moderate anemia before to the scheduled transfusion may impair their physical performance since thalassemia causes low hemoglobin levels, resulting in weariness and overall weakness, according to the patient.

Table4-2-3.Quality of Life related to Emotional Functioning

The mean score and standard deviations of the items reveals that adolescent are experienced poor emotional function as indicated by low mean scores among all items except, the items number (1) the responses were moderate emotional function as indicated by moderate level of mean.

This findings in the same line with study that carried out by (Mahmoud et al., 2019) who found that the highest percentages of those Adolescents had low QOL in relation to emotional.

Table4-2-4:Overall Quality of Life related to Emotional Functioning among Hemoglobinopathies Adolescent

The analysis of emotional function related to quality of life was demonstrate that highest percentage of adolescent are experienced moderate with average of *M* ± *SD=9.49±6.55*.

This findings in contrast with (Shaker and Nasir, 2013) who carried out a study about determination of quality of life for thalassemic adolescent and illustrated that the overall emotional QoL is (pass). This may be due to thalassemic adolescents feel different from their counterparts and develop a negative feelings about their life and their future, this bad feeling may come from the negative impact of the treatment and the disease itself, and the concern of overall health status or indications of recent deterioration in health.

Also , study to evaluate the health-related quality of life (HRQoL), muscular strength and pain in Adolescents with b-thalassemia major show that decrease in emotional domain (Ismail et al., 2018).

More than half of Adolescents studied with thalassemia had a decent quality of life in terms of emotional functioning, such as fear and anger, as described by the Adolescents and their caregivers. (Pakbaz et al., 2005) observed that emotional functioning is one of the weaker domains among thalassemia youngsters because they experience anxiety and concerns over their health state.

Table4-2-5.Quality of Life related to Social Functioning

The mean score and standard deviations of the items reveals that adolescent are experienced moderate social function as indicated by moderate mean scores among all items except, the items number (1 and 5) the responses were poor social function as indicated by low level of mean.

Table4-2-6:Overall Quality of Life related to Social Functioning among Hemoglobinopathies Adolescent.

The analysis of social functioning related to quality of life was demonstrate that highest percentage of adolescent are experienced moderate with average of $M \pm SD=10.66\pm5.98$.

This is consistent with the findings of Wahyuni et al., (2011), who conducted a cross-sectional study and found that the quality of life of thalassemic Adolescents was significantly lower than that of the control group. This may be due to, the thalassemia disease has different effect on Adolescents life.

Table4-2-6.Quality of Life related to School Functioning

The mean score and standard deviations of the items reveals that adolescent are experienced moderate school function as indicated by moderate mean scores among all items.

This findings in the line with study that carried out by (Ismail et al., 2013) entitled Quality of Life Among Thalassaemia Adolescents, Adolescent and Their Caregivers , who their findings indicated that lower emotional function among adolescent.

According to Wahyuni and colleagues (2011), difficulties at school may be caused by numerous variables, including missing school days due to the disease's burden (e.g. acute complications, hospitalizations, ED visits) and the neurological effects of the condition. Because of their frequent hospital visits for blood transfusions and iron chelating therapy, Thalassemic Adolescents are also frequently absent from school.

From other point of view is that the school sector was the worst hit. Patients with thalassemia’s face similar difficulties in school as those described in the literature, including frequent hospitalizations, poor peer group adjustment, and disease-related difficulties.

Table4-2-8:Overall Quality of Life related to School Functioning among Hemoglobinopathies Adolescent

The analysis of school functioning related to quality of life was demonstrate that highest percentage of adolescent are experienced moderate with average of *M* ± *SD=10.91±5.81*.

School functioning is lower in teenagers than in Adolescents under the age of 12 years, according to a study titled Health related quality of life in Malaysian Adolescents with thalassemia. Because of the cumulative effects of the disease and its treatment, this could be the case.

Patients with thalassemia are frequently unable to attend school due to the necessity of receiving regular blood transfusions in the hospital. The illness has a direct impact on their overall health. Emotional functioning (59.92 16.83 points), followed by social functioning (59.59 15.31 points) were the lowest psychosocial measures in a similar study (78.00 13.92 points) (Ismail et al. 2013).

Table4-2-9:Overall Quality of Life related among Hemoglobinopathies Adolescent

The analysis of quality of life among Hemoglobinopathies adolescents was demonstrate that more than half are experienced moderate quality of life with average of $M \pm SD=47.63\pm24.38$. this finding in the same line with (Hakeem et al., 2018) who reported that lower physical, social, emotional, school functioning, and total QoL scores compared with their matched healthy peers. This due to the thalassemia disease considered chronic disease that has major impact of different on Adolescents life like social relationship.

Table4.3.Health Behaviour among Hemoglobinopathies Adolescent

The mean score and standard deviations of the items reveals that adolescent are experienced poor level ($M> 2$) as indicated by low level of mean scores to moderate level ($M=2$-3) as described by moderate mean scores of the health behaviour at items of the scale. According to (Behdani et al., 2015) who conducted a study to evaluate psychological aspects and Quality of Life (QOL) in Iranian Adolescents and adolescents with thalassemia major compared to control group, their results showed that Also, the level of behavioral problems in these Adolescents is at a lower level compared to the control group of healthy subjects

Also, a study that carried out by (Siddiqui et al., 2014) This study found that many of these patients suffered from sadness, anger and loneliness. This might be due to the chronicity of the disease state that influences patient's recreational activities, capabilities and peer as well as family relationships, culminating in anxiety, secluded behavior and depression.

Table4-3-2:Overall Health Behaviour among Hemoglobinopathies Adolescent

Findings demonstrated that the (49%) of Hemoglobinopathies adolescents were poor health behaviour with average of $M \pm SD=94.0\pm28.74$.

This finding in parallel with (Khani et al., 2012) who carried out a cross-sectional descriptive study was conducted study , and reported that Due to distress from both the illness itself and the need for iron chelation, thalassaemia subjects frequently display maladaptive coping strategies and high levels of anxiety with psychosocial dysfunction . The probability of behavioural abnormality in Adolescents with TM has been estimated to be 1.6-fold higher than that in healthy Adolescents .

Behavioral difficulties were found in 32 percent of patients in a study conducted by (Hongally et al., 2012) to analyze the behavioral problems in multi-transfused thalassemic youngsters and psychosocial factors impacting them. Younger Adolescents, those with poor academic performance, were found to have higher CBCL scores.

4.4. Differences in Quality of Life and Health Behaviour with regards Adolescents Information

Table 4-4-1: Differences in QoL and Health Behaviour with regard Adolescents Age (*n=200*)

Quality of life differed significantly across age groups (p0.05), although there were no significant changes in health behavior among teenagers with hemoglobinopathies ($p>0.05$).

There was no statistically significant difference in the quality of life between men and women in any of the quality of life characteristics studied by

Jeesh and colleagues (2018). Patients and caregivers' knowledge, attitude, and practice affect quality of life among Thalassemia major patients in Damascus-Syrian Arab, according to research conducted by (Jeesh et al., 2018).

The researcher point of view this may have been because the studied Adolescents got used to the presence of the chronic disease while their ages advanced.

Table 4-4-2: Differences in QoL and Health Behaviour with regard Adolescents Gender (*n=200*)

Health behavior among hemoglobinopathies teenagers did not show any significant differences in gender (p>0.05), however there were significant disparities in quality of life among men and women (p0.05).

Adolescents in Abadan City who had beta-type majors were compared to their classmates, and the results showed that their QOL was much lower than that of their contemporaries without the beta-type majors. and showed that there was no correlation between quality of life and gender.

There were differences between men and women in terms of the four domains of quality of life, with men scoring higher than women in the psychological and environmental domains, while women scored higher than men statistically in social relations (71.222.2 compared with 66.126.2) (p=0.04), suggesting that men have a higher quality of life than women, according to (Jeesh et al. 2018), who reported that.

Also in other point of view this discrepancy may be attributed to a variety of factors, including race, age, and other factors. Choices about where and how to make clinical decisions should be made locally in each region.

Adolescent with thalassemia have a poorer QOL and a higher rate of depression than the general population as a result of the disease's progression and complications. These Adolescents also have fewer behavioral issues than healthy controls.

Table 4-4-3: Differences in QoL and Health Behaviour with regard Adolescents Education Level (*n=200*)

For hemoglobinopathies teenagers, there were no significant variations in quality of life and health behavior (p>0.05).

While the results of this study (Hongally et al., 2012) on behavioral problems and psychosocial factors in multi-transfused thalassemic Adolescents are incongruous with the latter's findings, the former found that statistically significant increases in CBCL scores were associated with Adolescents older than 7 years of age.

In the same context (Shawkat et al., 2019) who carried out a study about Evaluating Health-Related Quality of Life (HRQoL) in Iraqi Adult and Pediatric Patients with Beta-Thalassemia Major Using Two Different Iron Chelation Therapies and found that, while the educational level is correlated directly with QoL in univariate analysis.

In regarding to researcher point of view this may have been due to that education helps thalassemia Adolescents have suitable academic performance that makes them depend on themselves and socially live without problems.

Table 4-4-4: Differences in QoL and Health Behaviour with regard Adolescents Number of Sibling (*n=200*)

Hemoglobinopathies in teenagers and the number of siblings had no significant impact on quality of life or health behaviors, according to the findings (p-value=>0.05).

These findings are in line with (AL-Mosooi, 2011) who carried out a study and illustrated that Affected brother number ,which has no significant association with (QOL domains)

They found that thalassemia Adolescents have significantly lower quality of life than their non-thalassemia siblings (Wahyuni et al., 2011). The following were found in comparisons of the thalassemia and control groups in terms of quality of life assessments: The physical function score is 53.1, however the overall score is 71.5.

Table 4-4-5: Differences in QoL and Health Behaviour with regard Adolescents Order of Adolescents (*n=200*)

Findings showed that teenagers with hemoglobinopathies in their family had no significant variations in quality of life and health behavior (p-value=>0.05).

This contradicts the conclusions of (Wakimizu et al., 2020) who did a study. entitled Quality of life and associated factors in siblings of Adolescents with severe motor and intellectual disabilities: A cross-sectional study, and their results illustrated that association between quality of life and Adolescents order in the family.

Table 4-4-6: Differences in QoL and Health Behaviour with regard Disease Related Relives (*n=200*)

There were significant changes in health behaviors and health-related quality of life among adolescents with hemoglobinopathies, according to the findings (p0.05).

This findings congruent with (AL-Mosooi, 2011) who reported that At last the Family history has significant association with all quality of life domain. This may be due to family history playing a major role in the transmission of disease that led to effects on all aspects of life.

Table 4-4-7: Differences in QoL and Health Behaviour with regard Transfusion Therapy (*n=200*)

Among adolescents with hemoglobinopathies, the results showed no differences in health-related quality of life or behavior (p-value=>0.05).

This contrasts with the findings of (Bhattacharyya et al., 2019), who conducted a study. about a comparative study of temperamental, behavioral, and cognitive changes in thalassemia major, thalassemia minor, and normal population and their result indicated that number of blood transfusions has emerged up one of the significant predictors, and the difference is significant in TM group which for obvious reason required more blood transfusions at p-value=<0.001.

In the same direction, (Musa, 2017) who conducted a study and their results indicated that, there were significant associations in domains of quality of life with regard to frequency of blood transfusion.

Table 4-4-8: Differences in QoL and Health Behaviour with regard Iron Chelation Therapy (*n=200*)

Iron chelation therapy did not appear to affect teenagers with hemoglobinopathies' quality of life or health habits (p>0.05), according to the findings.

This findings incongruent with (Shafie et al., 2020) who carried out a study about who reported that there was significant correlational between HRQoL and Iron Chelation Therapy.

However, current study results were dissimilarity to Haghpanah et al. (2013), who discovered that Adolescents who were strict compliant with chelation treatment had high quality of life score. In addition, Goulas et al. (2012), mentioned that Adolescents receiving chelation therapy had lower self-esteem and worse physical scores.

Chelation treatment, which has been shown to increase survival and quality of life in transfused patients, may help to clarify this .

Table 4-4-9: Differences in QoL and Health Behaviour with regard Number of Transfusion Therapy (*n=200*)

When it came to the quality of life and health behavior, there were no significant variations in the number of transfusion therapy among adolescents with hemoglobinopathies.

This information (Ahmed Khalil et al., 2019) A recent study found that the QOL of those who complied with blood transfusions was low in the majority of the sample. Many health issues, including exhaustion, weakness, and impaired intellectual awareness can be explained by the fact that low hemoglobin levels cause these symptoms (Hockenberry et al., 2013).

The results of this study were congruent with Thavorncharoensap et al. (2010), their study clarified that for those who transfused by blood, there were positive relation which was significantly between Adolescents who received blood transfusion and their low health related quality of life, it might be clarified by Adolescents who receiving blood through 3 months before the health-related quality of life estimation who had minimum level of hemoglobin before transfusion. Also, Baraz et al. (2015), revealed the presence of negative relationship between Adolescents`s quality of life in addition to the frequency of blood transfusions.

4.5.Correlation between Quality of Life and Health Behaviour with regard to Hemoglobinopathies Adolescents their Parents Socio-Demographic Information

Table4-5-1:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Fathers Occupation

The quality of life of adolescents with hemoglobinopathies was shown to be significantly correlated (positively) with the occupation of their fathers (r=0.155; p=0.028).

According to (AlJabiri et al., 2015), who conducted a study about Assessment Quality of Life for Thalassemia Patients at Thalassemia Center, there was a correlation between the socio-demographic characteristics of nurses' age, gender, marital status, occupation, and blood group, as well as their effect on patient quality of life.

Table4-5-2:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Mothers Occupation

Findings exhibit there were significant correlation (negative) between health behaviour of hemoglobinopathies adolescents and mothers occupation (*r= -0.243; p-value=0.001*).

This research supports the conclusions of (Radha et al., 2015), who conducted a study. about Burden of Caregivers Care for Adolescents with Thalassemia at Babylon Adolescents and Maternity Teaching Hospital / Babylon Governorate and reported that there is signification relation between occupations of care givers with psychology burden at p≤ o.5 .

Table4-5-3:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Fathers Education

No link was found between the quality of life and health behavior of hemoglobinopathies teenagers and their fathers' education (p>0.05).

This study's findings are consistent with those of (Sadati and Tamaddoni, 2015), who found no connection between the well-being of adolescents and the educational attainment of their fathers.

Table4-5-4:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Mothers Education

The quality of life of adolescents with hemoglobinopathies was found to be significantly correlated (positively) with the education of their mothers (r= -0.161; p=0.022).

This findings in the same line with (Ahmed Khalil et al., 2019) who found that the correlation coefficient between Adolescents`s quality of life and mothers' education.

Table4-5-5:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Family Type

Findings exhibit there were significant correlation (positive) between health behaviour of hemoglobinopathies adolescents with regards family type (*r= -0.143; p=0.044*).

This findings in contrast with study that carried out by (Elsoudy et al., 2022) who reported that there is no relation between Adolescents who lived in extended homes and Adolescents who lived in nuclear homes in relation to compliance of Adolescents to their therapeutic regimen . It may be related to presence of relatives with Adolescents who lived in extended homes; who can caring for Adolescents, and helping mothers in giving care for their Adolescents; these type of homes acts as supportive family members

Table4-5-6:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Residences

Residents of hemoglobinopathies teenagers were not found to have a significant impact on their well-being (p>0.05).

They found similar things in a study done in Amirkola, Iran by (Sadati and Tamaddoni, 2015) on the quality of life in patients with beta-thalassemia major, and their results showed that there was no significant difference between rural and urban patients' quality of life when comparing the questionnaire.

A study conducted in Shahrekord by (Alavi and colleagues, 2007) found that in rural areas, Adolescents with substantial thalassemia had lower quality of life than those who resided in urban areas. While life expectancy differs slightly between north and south Iranians, access to health care facilities is less likely in the north and more common in south.

Table4-5-7:Correlation between QoL and Health Behaviour with regard to Hemoglobinopathies Adolescents their Family Economic

The results show a strong link (positive correlation) between hemoglobinopathies teenagers' quality of life (r=0.252; p=0.000) and health behavior (r=0.188; p=0.008) in terms of their parents' economic situation.

This findings go along with (Sinlapamongkolkul and Surapolchai, 2020) who carried out a study about Health-Related Quality of Life in Thai Adolescents with Thalassemia as Evaluated byPedsQL and EQ-5D-Y: A Single-Center Experience and demonstrated that significant differences between household income and PedsQL

In the same direction (Thiyagarajan et al., 2019) conducted a study about Assessing the role of family well-being on the quality of life of Indian Adolescents with thalassemia , and found that income have a direct influence on the Adolescents’s quality of life

Table 4-5.8 Correlation Between Quality of Life and Health Behaviour among Hemoglobinopathies Adolescents (*n=200*)

Findings show that adolescents with hemoglobinopathies had a favorable connection (r=0.260; p=0.000) between their health behaviors and their quality of life.

Halimah, Allendekania, and Waluyanti (2016) found that teenage thalassemia boys were more likely to suffer from behavioral issues like anxiety, social anxiety, and attention deficit disorder. However, this does not rule out the possibility of behavioral issues in adolescent girls as well. Other risk factors include ethnicity, multiple transfusions, maternal education, and the engagement of next of kin and relatives.

The researcher claims that thalassemia sufferers have a high number of emotions and behaviors that lower their self-esteem. In Aceh, Adolescents with thalassemian were found to have an abnormally high percentage of emotional and behavioral traits. A Adolescents's gender, age, and parent's work all play a role in how emotional and behavioral characteristics of thalassemia-affected Adolescents are described. This is in line with the findings of Aji et al. (2009), who found that people with thalassemia had a lower quality of life due to psychological and social changes. Adolescents with thalassemia require regular blood transfusions for the rest of their lives, and the associated medical expenses can be quite high. For both patients and their loved ones, long-term treatment can have a profound impact on their lives. For patients and their families alike, long-term treatment can have a profound effect on every part of life .

Chronic diseases are long-term, debilitating conditions that limit an individual's ability to choose their own destiny. Many chronic diseases necessitate significant lifestyle changes for their sufferers. The patient's and their family's coping mechanisms might be strained during periods of disease exacerbation and remission, as well as changes in health. As a result, changes in routine, such as taking on the role of a caretaker, changes in sexual life, social relationships and material life, all affect the psychological response to chronic illnesses. Adaptation is a necessary part of the treatment of chronic disorders. Depression, denial, low self-esteem, addiction, and family discord can all be symptoms of inadequate adaptation. Patients with the chronic condition thalassemia are subjected to both physical and psychological hardships. Due to improvements in the disease's course and a growing number of patients reaching maturity, psychological and social issues are on the rise. Patients with thalassemia major have a variety of challenges, just like those with other chronic illnesses. Mental health is often overlooked in the race to slow the progression of the disease.

6.1 Conclusions:

1. The highest percentage of parents were employed, most of parents graduate from institute and college graduated, Most of families were nuclear families, living in rural residents, and more than half slightly sufficient economic status. Less than half of adolescent have poor health behavior.

2. More than half of adolescent have moderate quality of life.

3. Less than half of adolescent have poor health behaviors.

6.2 Recommendations:

In the light of the results of the present study, the following recommendations are suggested:

1-Health educational programs about thalassemia disease and nursing care are essential for mothers and adolescents with beta thalassemia .

2-Beta thalassemia major education should be maintained through a multidisciplinary approach over a prolonged outpatient, inpatient, and community education service plan for mothers and adolescents with beta thalassemia .

3-Rehabilitation programs should be held for the adolescents with thalassemia and their caregivers to teach them stress management technique as progressive relaxation techniques, meditation and mindfulness.

4-Further studies with larger sample size or multicenter studies with longer period of follow-up.

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Appendix (C)

Administrative Arrangement

Ministry of Higher Education
and Scientific Research
وزارة التعليم العالي والبحث العلمي - جمهورية العراق
University of Babylon
College of Nursing
جامعة بابل
كلية التمريض
لجنة الدراسات العليا

Ref. No. :
Date: /

العدد : ٨٧٢
التاريخ : ١٧ / ٣ / ٢٠٢١

الدراسات العليا
جامعة بابل - كلية التمريض

الى / دائرة صحة كربلاء / مركز التدريب والتنمية البشرية
م/ تسهيل مهمة

تحية طيبة :
يطيب لنا حسن التواصل معكم ويرجى تفضلكم بتسهيل مهمة طالبة الدكتوراه (شهزنان حسن بدر عباس) لغرض جمع عينة دراسة الدكتوراه والخاصة بالبحث الموسوم :

تاثير اعتلال الهيموغلوبين طويل الامد على جودة الحياة وسلوك المراهقين في مركز امراض الدم في مدينة كربلاء.

Long term Impact of Hemoglobinopaties upon Quality of Life and Behaviors of Adolescents in Blood Diseases Center in Karbala City.

مع الاحترام ...

ا.م.د. ماهر خضير هاشم
العميد وكالة
٢٠٢١/٣/ ١٦

صورة عنه الى //
- مكتب السيد العميد للتفضل بالاطلاع مع الاحترام
- لجنة الدراسات العليا
- الصادرة

E-mail:nursing@uobabylon.edu.iq
STARS
07711632208 وطني
009647711632208 المكتب
www.uobabylon.edu.iq

جمهورية العراق

oly karbala governarate
arbala health directorate

محافظة كربلاء المقدسة
دائرة صحة كربلاء المقدسة
مستشفى كربلاء التعليمي للأطفال

العدد/ ٢٦٧٢
التاريخ / ٢٩ / ٥ / ٢٠٢١

الـى / دائرة صحة كربلاء المقدسة / مركز التدريب والتنمية البشرية / شعبة إدارة المعرفة / وحدة البحوث

م / عدم ممانعة

السلام عليكم ٠٠٠٠
كتابكم ذي العدد ٦٤٩ في ٢٠٢١/٥/٢٠
نود إعلامكم بعدم ممانعة مستشفانا من تسهيل مهمة طالبة الدكتوراه (شهزنان حسن بدر عباس) لغرض انجاز بحثها الموسو
(تأثير اعتلال الهيموغلوبين طويل الامد على جودة الحياة وسلوك المراهقين في مر
امراض الدم في مدينة كربلاء) وتكون الدكتورة (اسراء مصطفى الموسوي
المشرف العملي على البحث .
مع التقدير.

الدكتور
احمد توفيق عباس
مدير مستشفى كربلاء التعليمي للأطفال
٢٩ / ٥ / ٢٠٢١

دائرة صحة كربلاء المقدسة
مستشفى كربلاء للاطفال
الصادرة
العدد
التاريخ
/ / ٢٠١

صورة منه الى //
- وحدة التدريب والتطوير / الاضبارة العامة

ايمن//٥/٢٩

University of Babylon
College of Nursing
Research Ethics Committee

جامعة بابل
كلية التمريض
لجنة اخلاقيات البحث العلمي

Issue No: 39
Date: 16/03 /2021

Approval Letter

To,
Shahznan Hasan Badr

The Research Ethics committee at the **University of Babylon, College of Nursing** has reviewed and discussed your application to conduct the research study entitled "**Long-term Impact of Hemoglobinopathies upon Quality of Life and Behaviors of Adolescents in Blood Diseases Center in Karbala City.**"

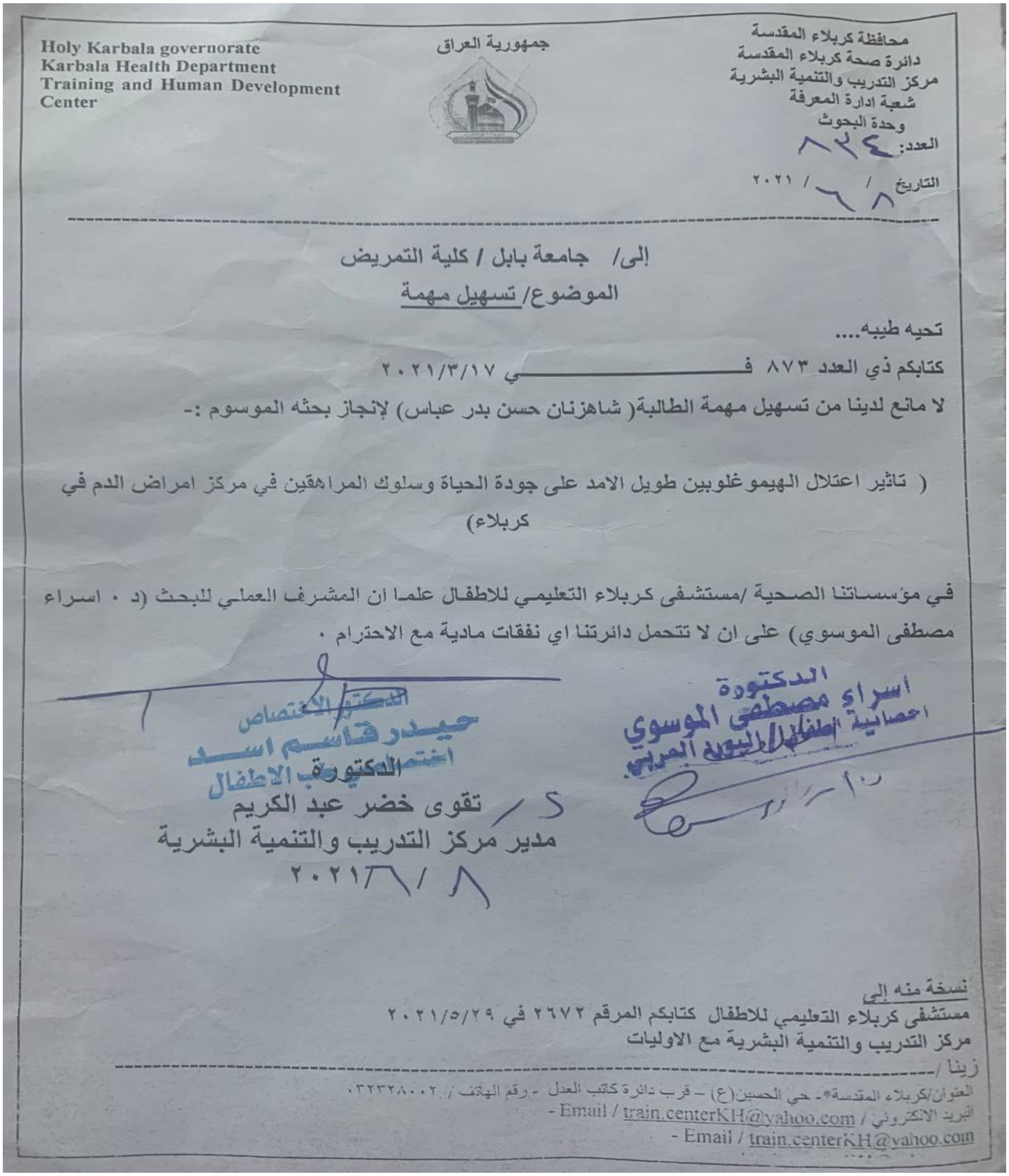
The Following documents have been reviewed and approved:

1. Research protocol
2. Research instrument/s
3. Participant informed consent

Committee Decision:

The committee approves the study to be conducted in the presented form. The Research Ethics committee expects to be informed about any changes occurring during the study, any revision in the protocol and participant informed consent.

Prof. Dr. Salma K. Jehad
Chair Committee
College of Nursing
Research Ethical Committee
16 / 3 /2021



Holy Karbala governorate
Karbala Health Department
Training and Human Development Center

جمهورية العراق

محافظة كربلاء المقدسة
دائرة صحة كربلاء المقدسة
مركز التدريب والتنمية البشرية
شعبة ادارة المعرفة
وحدة البحوث

العدد: ٨٢٤

التاريخ ٨ / ٦ / ٢٠٢١

إلى/ جامعة بابل / كلية التمريض

الموضوع/ تسهيل مهمة

تحيه طيبه....

كتابكم ذي العدد ٨٧٣ في ٢٠٢١/٣/١٧

لا مانع لدينا من تسهيل مهمة الطالبة(شاهزنان حسن بدر عباس) لإنجاز بحثه الموسوم :-

(تاثير اعتلال الهيموغلوبين طويل الامد على جودة الحياة وسلوك المراهقين في مركز امراض الدم في كربلاء)

في مؤسساتنا الصحية /مستشفى كربلاء التعليمي للاطفال علما ان المشرف العملي للبحث (د ٠ اسراء مصطفى الموسوي) على ان لا تتحمل دائرتنا اي نفقات مادية مع الاحترام ٠

الدكتور الاختصاص
حيدر قاسم اسد
اختصاص طب الاطفال

الدكتورة
أسراء مصطفى الموسوي
اخصائية طب الاطفال البورد العربي

الدكتورة
د / تقوى خضر عبد الكريم
مدير مركز التدريب والتنمية البشرية
٨ / ٦ /٢٠٢١

نسخة منه إلى
مستشفى كربلاء التعليمي للاطفال كتابكم المرقم ٢٦٧٢ في ٢٠٢١/٥/٢٩
مركز التدريب والتنمية البشرية مع الاوليات
زينا /

العنوان/كربلاء المقدسة*- حي الحسين(ع) – قرب دائرة كاتب العدل - رقم الهاتف / ٠٣٢٣٢٨٠٠٢
البريد الالكتروني / train.centerKH@yahoo.com - Email /
- Email / train.centerKH@yahoo.com

Appendix (B)

List of Experts

Appendix (B) List of Experts

ت	أسم الخبير	اللقب العلمي	مكان العمل	التخصص	سنوات الخبرة
١.	أ. د. عفيفة رضا عزيز	أستاذ	جامعة بغداد- كلية التمريض	تمريض الاطفال	٤0
٢.	أ.د أمين عجيل الياسري	أستاذ	جامعة بابل –كلية التمريض	تمريض صحة الأسرة والمجتمع	٣٨
٣.	أ..د .نهاد محمد الدوري	أستاذ مساعد	جامعة بابل-كلية التمريض	تمريض اطفال	٣٥
٤.	أ. د. حسين هادي عطية	أستاذ	جامعة بغداد- كلية التمريض	تمريض بالغين	٣٥
٥.	أ .د. سحر ادهم علي	أستاذ	جامعة بابل –كلية التمريض	تمريض بالغين	٣٠
٦.	أ.د وسام جبار قاسم	أستاذ	جامعة بغداد-كلية التمريض	تمريض صحة الأسرة والمجتمع	٢٥
٧.	ا.د خميس بندر عبيد	أستاذ	جامعة كربلاء – كلية التمريض	تمريض الأطفال	٢١
٨.	أ. م. د . مهدي عبد نعمة	أستاذ مساعد	جامعة بغداد- كلية التمريض	تمريض الاطفال	١٨
٩.	أ. م.د. حيدر حمزة الحدراوي	أستاذ مساعد	جامعة الكوفة-كلية التمريض	تمريض الصحة النفسية والعقلية	١٨
١٠.	أ.م.د ضياء كريم البياتي	أستاذ مساعد	جامعة العميد-كلية التمريض	تمريض بالغين	15
١١.	أ.م.د زيد وحيد عاجل	أستاذ مساعد	جامعة بغداد-كلية التمريض	تمريض الأطفال	١٢
١٢.	أ.م.د. صادق عبد الحسين حسن	أستاذ مساعد	جامعة بغداد-كلية التمريض	تمريض بالغين	١٠
١٣.	أ. م.د. رضا محمد عبد لفتة	استاذ مساعد	جامعة العميد- كلية التمريض	تمريض صحة الاسرة والمجتمع	٦

Appendix (A)

Questionnaires Format

Appendix (A)

Questionnaires in Arabic

(تأثير اعتلال الهيموغلوبين طويل الامد على جودة الحياة وسلوك المراهقين في مدينة كربلاء)

الجزء ١: المعلومات الديموغرافية

١.الجنس ذكر ☐ أنثى ☐

٢.العمر ☐

٣.المستوى التعليمي لايقرء ويكتب ☐ يقرء ويكتب ☐ خريج ابتدائيه ☐
خريج ثانوية ☐

٤.عدد الاخوة والاخوات ☐

٥.ترتيب الطفل في العائلة (١-٣) ☐ (٤-٧) ☐ (٧ او اكثر) ☐

الجزء ٢: معلومات حول تاريخ المرض

١.حدوث المرض ☐

٢. أحد أقاربك مصاب بالمرض نعم ☐ لا ☐

إذا كان الجواب نعم ، اذكر العلاقة الدرجة الاولى(الاب والام والاخوة والاخوات) ☐ الدرجة الثانية (الجد والجدة) ☐ الدرجة الثالثة (الاعمام والاخوال) ☐

3.نقل الدم منتظم ☐ غير منتظم ☐

4.العلاج باستخلاب الحديد منتظم ☐ غير منتظم ☐

5.عدد عمليات نقل الدم كل شهر مرة واحدة ☐ مرتين ☐

الجزء ٢: الوضع الاجتماعي والاقتصادي للأسرة

١.وظيفة الوالدين:

مهنة الاب	مهنة الام
موظف ☐	موظفة ☐
اعمال حرة ☐	اعمال حرة ☐
متقاعد ☐	متقاعدة ☐

كاسب ☐ ربة بيت ☐

متوفي ☐ متوفية ☐

٢. مستوى تعليم الوالدين: مستوى التعليمي للاب مستوى التعليمي للام

لايقرء ولايكتب ☐ لاتقرء ولاتكتب ☐

يقرء ويكتب ☐ تقرء وتكتب ☐

خريج ابتدائية ☐ خريجة ابتدائية ☐

خريج ثانوية ☐ خريجة ثانوية ☐

خريج معهد او كلية ☐ خريجة معهد او كلية ☐

٣.نوع الاسرة الاسرة نووية (تتكون من الاب والام فقط) ☐ الاسرة ممتدة (تتكون من الاب والام والجد والجدة) ☐

٤.عنوان السكن مدينة ☐ الريف ☐

٥.عدد افراد العائلة ☐

٦.دخل العائلة لايكفي ☐ يكفي قليلا ☐ يكفي ☐

الجزء الثالث :جودة الحياة للمراهقين

الاداء البدني	ابدا (٠)	قليلا (١)	بعض الاحيان (٢)	غالبا (٣)	دائما (٤)
١.اواجه صعوبة في المشي اكثر من ١٠٠ متر					
٢ اواجه صعوبة في الركض					
٣. اواجه صعوبة عند ممارسة النشاطات والتمارين الرياضية					
٤. اواجه صعوبة عند رفع الاشياءالثقيلة					
٥. اواجه صعوبة عند الغسل					
٦. اواجه صعوبة عند القيام بالمهام الروتينية حول المنزل					
٧. اعاني من الالم واوجاع في جسمي					
٨. اشعر بفقدان الطاقة					
الاداء العاطفي					
١ انا اشعر بالخوف او التعب					

٢ انا اشعر بالحزن او الكئابة					
٣ انا اشعر بالغضب					
٤ انا لدي مشاكل عند النوم					
٥ انا قلق عما سيحدث لي					
الاداء الاجتماعي					
١ انا اجد صعوبة بالتعامل مع الاولاد					
٢. المراهقون الاخرون لايرغبون ان يكونوا اصدقائي					
٣. المراهقون الاخرون يغيضونني					
٤. انا لا استطيع القيام باشياء يمكن للمراهقون الاخرون القيام بها					
٥. اواجه صعوبة عند العب مع المراهقين الاخرين					
الاداء المدرسي					
١. اواجه صعوبة في التركيزفي الصف					
٢ انا لا اتذكر الاشياء					
٣.اواجه صعوبة عند اداء واجباتي المدرسية					
٤. اتغيب عن المدرسة					
٥.اتغيب عن المدرسة لكي اذهب الى الدكتور اوالمستشفى					

الجزء ٤: سلوكيات المراهقين

	لا مانع على الاطلاق (٠)	قليلا فقط (١)	الى حدا كبير (٢)	كثيرا جدا (٣)
١. أجد صعوبة في الجلوس				
٢. اتحمس بسهولة				
٣ .افعل اشياء بدون تفكير				
٤ .اواجه صعوبة في التركيز				
5.انا اتعصب بسهولة				
٦. أواجه مشكلة عند التعامل مع الكبار ، مثل مديري المدارس والمعلمين وأرباب العمل.				
٧. اتشاجر مع أفراد عائلتي.				
٨.أتشاجر مع أشخاص من خارج عائلتي.				
٩. مزاجي يتغير سريعا .				
١٠.اتعمد خرق القواعد أو عصيان التعليمات.				
١١.افعل اشياء خطيرة				
١٢ .اخاف ان افقد السيطرة اوان اكون عنيفا				
١٣.انا عنيد او قوي الارادة				
١٤.اسرق اشياء				
١٥. اكذب على الناس				
١٦. أتغيب عن المدرسة.				
١٧. أنا غير منظم				
١٨. أجد صعوبة في تكوين صداقات أو الاحتفاظ بها.				
١٩. من السهل تشتيت انتباهي بسبب الضوضاء.				
٢٠.أجد صعوبة في البقاء جالسا.				
٢١ .لدي مشاكل في انتظار دوري في الألعاب أو المواقف الجماعية.				
٢٢ . أنسى القيام بالأعمال المنزلية أو الأشياء الأخرى التي يُطلب مني القيام بها.				
٢٣.غالبًا ما ابوح في إجابات للأسئلة قبل اكتمالها.				

٢٤انا اتحدث كثيرا				
٢٥ . أشعر بالقلق والتوتر				
٢٦. انا ابكي بسهولة				
٢٧. أجد صعوبة في التعلم في المدرسة.				
٢٨. لدي مشاكل في الانتباه للتعليمات.				
٢٩. أقاطع الآخرين عندما يتحدثون.				
٣٠. أفقد الأشياء الضرورية لأداء واجباتي الصفية (على سبيل المثال ، قلم رصاص ، كتب).				
٣١. أنا لا أكمل واجبي الدراسي				
٣٢. اتعامل بعنف مع الحيوانات أو أشخاص.				
٣٣.أقسم أو أستخدم لغة سيئة.				
٣٤ . أنا أزعج الآخرين.				
٣٥. انا الاوم الاخرين على اخطائي				
٣٦. انا اضيع الوقت عندما ادرس				
٣٧ . معظم أصدقائي أصغر مني.				
٣٨. انا لدي مشاكل في تذكر ما قرات				
٣٩. أصدقائي يتضايقون مني				
٤٠ .أنا لا أتعايش مع أقراني من الجنس الآخر.				
٤١. انا لا اكمل واجباتي المنزلية				

Questionnaires in English

Part 1:Demographic information

1. Gender Male ☐ Female ☐

2.Age ☐

3.Adolescentss level of education Not read and ☐ Read and write ☐
Primary ☐ High School and ☐

4.Numer of sibling ☐

5.Adolescentss order in the family ☐ ☐ (7 or m ■

Part 2 : Information about disease history

1. Disease occurrence ☐

2.One of your relatives with disease yes ☐ No ☐

Lf the answer is Yes , mention the relationship

1. First Degree ☐ 2. Second Degree ☐ 3.Third Degree ☐

3.Transfusion therapy Regular ☐ Not regular ☐

4. Iron chelation therapy Regular ☐ Not regular ☐

5. Number of transfusion therapy / month Once ☐ Twice ☐

Part2: Social and Economic Status of the Family

1.Parents Job :

Father Job Mothers Job

Employee ☐	Employee ☐
Acts Free ☐	Acts Free ☐
Retired ☐	Retired ☐
Un employee ☐	House Wife ☐
Death ☐	Death ☐

2.Parents Education Level:

Fathers education level	Mothers education level
Not read and write ☐	Not read and write ☐
Read and Write ☐	Read and Write ☐
Primary School ☐	Primary School ☐
High School Graduate ☐	High School Graduate ☐
Institute or College ☐	Institute or College ☐

3.Family type

A. Nuclear Family ☐

B. Extended Family ☐

4.House address Rural ☐ Urban ☐

5.Family number ☐

6. Family Income Insufficient ☐ Slightly sufficient ☐
Sufficient and save ☐

Part 3:Quality of Life for Adolescent

Physical functioning	**Never (0)**	**Almost never(1)**	**Sometimes (2)**	**Often (3)**	**Almost always (4)**
1.It is hard for me to walk more than 100M					
2. It is hard for me to run					
3. It is hard for me to do					

sports activity or exercise					
4. It is hard for me to lift something heavy					
5. It is hard for me to take a bath or shower by myself					
6. It is hard for me to do chores around the house					
7. I hurt or ache					
8. I have low energy					
Emotional Functioning					
1. I feel afraid or scared					
2. I feel sad or blue					
3. I feel angry					
4. I have trouble sleeping					
5. I worry about what will happen to me					
Social Functioning					
1. I have trouble getting along with other Adolescentss					
2. Other teenagers do not want to be my friend					
3. Other teenagers tease me					
4. I cannot do things that other teenagers my age can do					
5. It is hard to keep up when I play with other teenagers					
School Functioning					

1. It is hard to pay attention in class					
2. I forget things					
3. I have trouble keeping up with my schoolwork					
4. I miss school because of not feeling well					
5. I miss school to go to the doctor or hospital					

Part 4: Behaviors for Adolescent

	Not At All (0)	**Just A Little (1)**	**Pretty Much (2)**	**Very Much (3)**
1.I have difficulty sitting still				
2. I get excited easily				
3. I say or do things without thinking.				
4. I have trouble concentrating.				
6. I get frustrated easily.				
7. I get angry easily.				
8. I get into trouble with adults, such as principals, teachers, employers. dangerous things without thinking what might happen.				
9. I argue with my family members.				
10. I argue with people outside my family.				
11. My moods change quickly.				
12. I purposely break rules or disobey instructions.				
13. I do dangerous things without thinking what might happen.				
14. I'm afraid of losing control or				

becoming violent.				
15. I am stubborn or strong-willed.				
16.I get into trouble with the police; I break laws.				
17. I steal things.				
18. I lie to people.				
19. I skip school; I am truant.				
20. I am disorganized.				
21. I have trouble making or keeping friends.				
22.My mind wanders easily; I daydream.				
23. I am easily distracted by noises.				
24. I have trouble staying seated.				
25. I have problems waiting my turn in games or group situations.				
26. I forget to do my chores or other things I'm asked to do.				
27. I often blurt out answers to questions before they are completed.				
28. I talk too much.				
29. I feel restless and nervous.				
30. I cry easily.				
31. I have difficulty learning in school.				
32. I have problems paying attention to instructions.				
33. I interrupt others when they are speaking.				
34. I lose things necessary to do my classwork (e.g., pencil, books).				
35. I don’t complete my classwork.				
36. I’m physically cruel to animals or people.				
37. I swear or use bad language.				
38. I disturb or annoy other people.				
39. I blame others for my own mistakes.				
40. I get into physical fights.				
41. I waste time when studying.				
42. Most of my friends are younger than I am.				

43. 1 have problems remembering what I read.				
44. My friends get annoyed with me.				
45. I don’t get along well with peers of the opposite sex.				
46. I don’t complete my homework.				
47. I make careless errors in my classwork or homework.				
48. My classwork or homework is sloppy.				

Appendix (D)

Linguistic Approval

الخلاصة

اعتلال الهيموغلوبين هي أكثر الاضطرابات أحادية الجين التي تهدد الحياة شيوعًا في العالم. الأسباب الأكثر شيوعًا لاعتلال الهيموغلوبين هي مرض فقر الدم المنجلي والثلاسيميا .

الهدف الرئيسي من هذه الدراسة هو تقييم جودة الحياة وسلوك المراهقين المصابين باعتلال الهيموغلوبين (الثلاسيميا وفقر الدم المنجلي).

اجريت دراسة مقطعية وصفية من الأول من تشرين الثاني/ ٢٠٢٠ الى الثالث من ابريل/ ٢٠٢٢ . شملت الدراسة (٢٠٠) مراهق في ردهات أمراض الدم الوراثية في مستشفى كربلاء التعليمي للأطفال. كانت عينةالدراسة عينة مناسبة (ملائمة)غير احتمالية .

تم تحديد صلاحية الاستبيان من خلال توزيعها على (١٣) خبيراً. أجريت الدراسة التجريبية على (٢٠) مراهق لتحديد مصداقية الاستبيان. تم تحليل البيانات من خلال الإحصاء الوصفي والاستنتاجي.

تتراوح أعمار غالبية المراهقين بين١٤-١٦عامًا. وكانت غالبية المراهقات من المدرسة الابتدائية ، ولديهن اخ واحد ، والتسلسل الثاني داخل أسرهن. كان نصفهم من الذين تلقوا علاجًا منتظمًا ، وخضعوا للعلاج بنقل الدم مرتين. كانت أعلى نسبة من الآباء يعملون ، ومعظم الآباء تخرجوا من المعاهد والكليات ، وكانت معظم العائلات من الأسر النووية ، وتعيش في سكان الريف ، وكان وضعهم الاقتصادي يكفي قليلاً. لوحظت السلوكيات الصحية السلبية لأقل من نصف المراهقين. ارتبطت نوعية حياة المراهقين المرتبطة باعتلال الهيموغلوبين بشكل إيجابي باحتلال والدهم. أظهر اعتلال الهيموغلوبين لدى المراهقين جودة الحياة والسلوكيات الصحية ارتباطًا قويًا (إيجابيًا).

استنتجت الدراسة إلى أن أقل من نصف المراهقين يعانون من سوء السلوك الصحي وأن أكثر من نصف المراهقين يتمتعون بنوعية حياة معتدلة ، بالاضافة الى ذلك تأثر نوعية الحياة والسلوكيات الصحية للمراهقين المصابين باعتلال الهيموغلوبين بعدة عوامل منها (وضيفة الوالدين ، والوضع الاجتماعي والاقتصادي لأسرهم) بالإضافة إلى ذلك ، استنتجت الدراسة إلى وجود تأثير معنوي للسلوك الصحي على نوعية حياة المراهقين المصابين باعتلال الهيموغلوبين.

أوصت الدراسة بأن المراهقين المصابين بالثلاسيميا بيتا وأمهاتهم يحتاجون إلى علاج شامل يجب الحفاظ على التعليم من خلال نهج متعدد التخصصات على مدى فترة طويلة من العيادات الخارجية ، والمرضى الداخليين ، وخطة خدمة التثقيف المجتمعي للمراهقين المصابين بيتا ثلاسيميا وأمهاتهم .

جمهورية العراق

وزارة التعليم العالي والبحث العلمي

جامعة بابل

كلية التمريض

تأثير أعتلال الهيموغلوبين طويل الامد على جودة الحياة وسلوك المراهقين في مركز امراض الدم في مدينة كربلاء.

اطروحة مقدمة من قبل

شهزنان حسن بدر

الى مجلس كلية التمريض، جامعة بابل كجزء من متطلبات نيل درجة الدكتوراه فلسفة في علوم التمريض

باشراف

أ.د. عبد المهدي عبد الرضا حسن

ايار/٢٠٢٢ ميلادي **شوال/ ١٤٤٤ هجري**